



THE GENTLE WAY THE ART OF PAEDIATRIC IMAGING





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IMAGING THE YOUNG PATIENT: AN INTRODUCTION

BY CATHERINE OWENS

e are delighted to be given this unique opportunity to share our working experience with you. As a community of imaging specialists, who spend our working lives imaging children, we are particularly enthusiastic to have this chance to show-case our particular knowledge and skills in trying to help all children receive the best possible medical care that is achievable. We do this in a variety of ways, and we hope this book will help to explain them, as well as guide you through understanding some of the challenges that we face in trying to achieve this goal.

For each of the last four years, the European Society of Radiology (ESR) has put together a book like this one to mark the International Day of Radiology (IDoR), celebrating the dedication, hard work and particular skills of a specific sub-group of radiologists. The ESR leadership and our partners at the Radiological Society of North America and the American College of Radiology hope to publicise and highlight the contribution that is made to each individual patient, via our teams of experienced and well trained imaging specialists, alongside nurses and other healthcare professionals that look after these patients every day. We are honoured that in its fourth year, the IDoR organisers have chosen paediatric radiology as an important field to highlight, and on November 8, 2015, we will celebrate the International Day of Radiology, illustrating and highlighting the work that paediatric radiologists undertake in their careers, performing the best imaging in children to obtain the best possible standards of care.

Paediatric imaging is a varied and very exciting medical subspecialty, partly due to the diverse range of imaging techniques that we have at our disposal, and particularly because we deal with the diverse developmental stages from foetal life (during pregnancy), through early childhood, to adolescence, that amounts to more than 18 years of life maturation and growth. As many different diseases occur at different ages, and children with the same disease require a different approach at different ages, it truly provides a wide spectrum of medicine, both diagnostic and therapeutic, and simultaneously both challenging and stimulating.

Many of the authors of the articles in this book are members and officials of the European Society of Paediatric Radiology (ESPR), which was founded in 1963 and is hence one of the oldest subspecialty societies in Europe, as well as of its sister society in North America, the Society for Paediatric Radiology (SPR). Over the last few decades, these societies and their counterparts in South America, Asia, Australasia and Africa, as well as the World Federation of Paediatric Imaging (WFPI), have become the functional backbone to support, encourage and enhance doctors licensed to practice medicine, whose main area of work is within paediatric imaging or image-guided intervention. Through the extended ESPR and SPR communities, paediatric imaging specialists are able to teach and learn from experienced colleagues, and share their challenging experiences with other specialists, in order to guide the successful future of a growing and very exciting and rewarding specialty. We have a close professional collaboration with the various adult radiology societies, in order to share our experiences within similar anatomic fields, in the growing and developing bodies of children. This helps the transition adolescents take into the world of adult medicine.

In the first part of this publication, we will outline the different imaging techniques that we use on a daily basis and highlight the need for specifically trained, highly experienced paediatric radiologists to image children. We aim to provide the best quality imaging within the shortest possible time using the lowest possible radiation dose. Many of our techniques involve ionising radiation, and children's developing tissues are more sensitive to effects of radiation, as they have a longer lifespan ahead of them in which to develop any potential radiation-induced side effects.

Within paediatric imaging, recent global initiatives to protect and try to homogenise and improve the standards of care for children include the ESR-led *EuroSafe Imaging* campaign and the SPR and ACR contributions for the Alliance for Radiation Safety in Pediatric Imaging *Image Gently* campaign. These organisations and the projects they coordinate attempt to address global barriers to the best possible paediatric imaging, allowing for the very different and varied facilities that can be provided around the world, and to promote education, best practice and appropriate imaging guidelines for the benefit of all children, particularly addressing those in developing and resource-poor areas.

In the second part of our publication, we outline several common and important medical conditions that

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affect children, and describe and highlight how we use the selection of imaging techniques at our disposal, to optimise the management of young patients. We aim to provide the best answers at each stage to guide the most appropriate and effective treatment available, for the most beneficial long-term outcome.

In order to continue to improve and enhance this most exciting and vital subspecialty, ongoing medical research is needed. However, performing clinical research within paediatric radiology is a major challenge, due to the relatively small number of children with specific diseases. The best way to address this is to involve several centres across different countries and even continents, to summate data, and to work closely with manufacturers, although there are obvious difficulties and conflicts in doing so. At the end of this publication, we summarise these difficulties and how they might be overcome.

We have also included a number of interviews with experienced and world renowned paediatric radiologists who share with you their daily practice, to give some personal insight into how we can move forward most effectively for the good of children everywhere.

We are proud and excited to share our expertise with you, our readers, to enable you to understand our daily practice and to share some of the fascinating diagnostic and therapeutic challenges that is our work, undertaken for our children. We will highlight the exciting developments that are in progress - many linked to the rapid expansion of the computer-driven twenty-first century, from which radiology benefits hugely - and how we translate this computer technology for the good of children's healthcare. We hope that you will enjoy reading this publication, as much as we have loved sharing our work with you. We trust that you will join us in celebration of the valuable and vital work done by paediatric imaging specialists in medical centres around the globe. We know that you will appreciate how we need to unite together to provide the best possible care for our children everywhere.



WHAT DOES A PAEDIATRIC RADIOLOGIST DO? ASPECTS OF EARLY PAEDIATRIC RADIOLOGY WHY IMAGING CHILDREN IS DIFFERENT TO IMAGING ADULTS IMAGE GENTLY®: A BIG VOICE FOR OUR LITTLE ONES RADIATION PROTECTION IN ACTION: EUROSAFE IMAGING THE IMPORTANCE OF A CHILD-FRIENDLY IMAGING ENVIRONMENT GLOBAL PAEDIATRIC RADIOLOGY



AN INTRODUCTION TO PAEDIATRIC IMAGING



WHAT DOES A PAEDIATRIC RADIOLOGIST DO?

BY KATHARINE HALLIDAY

paediatric radiologist is a specialist doctor who is skilled in performing and interpreting children's scans to make a diagnosis. The radiologist may also use images to carry out treatment. Their work is extremely varied as children come in a variety of sizes, from tiny premature babies to large adolescents. In fact, many paediatric radiologists also advise on the imaging of foetuses before they are born, using ultrasound or magnetic resonance imaging.

Children's radiology differs from the adult branch of the specialty in almost every respect. The diseases affecting children are often completely different; foetuses and young children may be affected by abnormalities of development, which require imaging to guide and plan treatment, and also to accurately diagnose genetic abnormalities within a family. Some diseases are only found in children; for instance, cancers such as Wilms tumour and neuroblastoma only occur in children, and other conditions are seen only in newborn or premature babies. Conditions which affect all ages may have different manifestations in younger patients. For instance, urinary tract infec-

tions in adults are usually not concerning, but the same infections in children can damage the growing kidneys. Medical management is aimed at protecting kidney function and growth, and imaging is required to monitor renal growth and look for scarring in the kidney.

The paediatric radiologist advises the paediatrician or surgeon which is the best sort of imaging to investigate a particular problem. Possible methods (modalities) include plain radiographs (x-rays), ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), fluoroscopy and nuclear medicine. The choice of modality for imaging children is influenced by several factors. First, of course, which technique is most likely to identify or rule out the abnormality suspected, based on knowledge of the disease itself. But children are more sensitive to radiation than adults and therefore x-rays, fluoroscopy and CT (which all use ionising radiation) are used as little as possible. Wherever feasible, ultrasound and MRI are used as an alternative. When ionising radiation is used, the paediatric radiologist works with the technologists to ensure that the examination is performed in such a gist to make a diagnosis.

The choice of imaging test may also be influenced by the patient's ability to cooperate with the examination. A child may be unable to hold their breath or lie still for the required length of time. Some examinations require the insertion of catheters (tubes) into the bladder or rectum which can be very frightening to the child. Paediatric radiologists and technologists work together to provide a reassuring, child friendly environment to minimise the stress of the situation for the patient and their family and to enable the child to cooperate, so that the maximum information possible can be gained from the examination. Considerable skill and experience is required to obtain the best possible image in the shortest possible time. Compromises are often made; the paediatric radiologist or sonographer becomes adept at getting the fewest ultrasound images of a mobile toddler that are just enough to make a confident diagnosis. Any experience in the x-ray department which makes a child anxious can make future examinations even more difficult to per-



way as to minimise radiation exposure whilst ensuring the image is of a good enough quality for the radiolo-

form, so the specialist expertise of the paediatric radiologist is invaluable in knowing what information is crucial

The radiologist's role is to evaluate each child, weigh up the balance of risk versus the benefit of each test and design the best imaging strategy so that the maximum information is obtained and the child and their family remain as comfortable, relaxed and reassured as possible.

While many radiologists may have only limited interaction with adult patients, as they can work largely from a computer in an office, this is not true of the paediatric radiologist. Communication skills are therefore paramount. It is vital to have a realistic idea of what is possible for each patient, and the radiologist must be able to communicate clearly with their patient in a way which is appropriate to the child's age. In addition to the patient, the radiologist must also communicate with the child's carers, who usually have different but equally important needs. The parents' or carers' understanding and cooperation, not to mention consent, is vital for success. Carers at

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each imaging examination will often be eager to learn the result immediately and the radiologist must work closely with clinical colleagues and be sensitive to the context of their interaction, taking into consideration what the family already know of their child's condition and the environment and support available to them. For instance, it is best to discuss the possible diagnosis of a cancer, which has significant implications for the child and family, in a quiet space with appropriate support for the family. It is equally important to provide accurate information when asked directly by a parent or carer.

In addition, it is also important to understand when a child may be old enough to make decisions about their own care independent from their carers and to respect their confidentiality in these cases. This can be a very difficult area and requires skill and sensitivity.

The results of any investigations must then be communicated clearly to the clinical team responsible for the treatment of the patient, highlighting the advantages and limitations of the findings. This often takes place in a multidisciplinary team meeting where all the health professionals involved in the patient's care discuss each case in depth and work out a management

plan, often guided by the imaging provided by the paediatric radiologist.

So how does one become a paediatric radiologist? This can vary depending on which part of the world you are in, but all paediatric radiologists have usually undergone basic medical training and have completed at least one year (and often several years) as a clinical hospital doctor. After that, general radiology training is undertaken followed by a minimum of one or two years' paediatric radiology training before taking up a post as a specialist. Paediatric radiologists usually work in children's hospitals or children's units in general hospitals. They also often provide specialist input to smaller, less specialised hospitals from a distance, taking advantage of the advances in computerised image transfer that have been made in recent years.

The paediatric radiologist therefore has an intensely varied and stimulating role. Each day is different to the last and we are continually tested and stretched in terms of scientific knowledge, communication skills and time management. Energy, pragmatism, humility, empathy and a sense of humour are vital characteristics for this job, but if you have these and enjoy a challenge, the rewards are immense.

"The paediatric radiologist has an intensely varied and stimulating role. Each day is different to the last and we are continually tested and stretched in terms of scientific knowledge, communication skills and time management. Energy, pragmatism, humility, empathy and a sense of humour are vital characteristics for this job, but if you have these and enjoy a challenge, the rewards are immense."

Katharine Halliday



ASPECTS OF EARLY PAEDIATRIC RADIOLOGY

BY ADRIAN THOMAS

THE EARLY YEARS

hortly after x-rays were discovered by Wilhelm Conrad Röntgen on November 8, 1895, the first radiographs of children were made. Röntgen had sent details of his discovery to physicists throughout the world, including Arthur Schuster of Manchester, and the discovery was greeted with astonishment by scientists and general public alike. Schuster had been sent an envelope containing photographs, among them one showing the outlines of a hand with the bones clearly marked, and a thin pamphlet titled Ueber eine neue Art von Strahlen, by W.C. Röntgen. Schuster translated this 'first communication', which he sent to the scientific journal Nature in 1896¹. The apparatus that Röntgen had used to make his astonishing discovery was available in any physics laboratory, and so it was straightforward for others to replicate his work. This is exactly what Arthur Schuster

did, and one of his early subjects was his own young son who had injured himself. I met his daughter Norah many years later, and she recalled how absolutely terrified her brother was by the x-ray apparatus!

Some of the earliest radiographic examinations were of infants and children and the early history was described well by the great American radiologist John Caffey in the preface to the first edition of his classic book *Pediatric X-ray Diagnosis* in 1945². It might be thought that thereafter, because of the relatively low power of the early apparatus, and the small size of children compared to adults, the new x-ray technique would be commonly used in children. In fact this was far from the case, and John Caffey quotes Dr. Theodor Escherich from Graz in Austria, who had pointed out that, even by 1898, radiography was not being used as commonly in young patients as in adults. However, a Röntgen laboratory specifically for children had been established in Graz by 1897, and this was probably the first of its kind. The first textbook devoted to radiology in any language was written by Thomas Morgan Rotch in 1910. Rotch was Professor of Pediatrics at Harvard University and a pioneer of neonatal care. The book is fascinating and gives a brilliant account of living anatomy and pathology with many high quality radiographs. Rotch stressed the importance of mastering normal appearances before interpreting the abnormal, and the book was based on his experience of Boston Children's Hospital. During the subsequent decades, a number of books appeared in German, however it is quite surprising that when Caffey published his book in 1945 that there had been no previous textbook in English. Writing in 1955, Caffey stated that paediatric radiology was becoming more widely used and the accuracy had greatly improved. He was pleased that the first two editions of his book were stimulating growth and interest in paediatric radiology. By this third edition there had been many new changes in paediatric radiology, including many developments that were quite invasive compared to our modern techniques. Caffey was particularly keen to point out that in radiology practice and teaching they had not talked about the limitations of paediatric radiology. He felt that the greatest value of radiology was in suggesting a line of investigation so that a conclusive diagnosis could be made. The radiological findings were often seriously misleading when the radiologist was not familiar with normal variations and the limitations of the technique, and this is as true today as it was 60 years ago in 1955.

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As new techniques were introduced they were applied to infants and children. However, there were specific issues that apply to children, in particular related to the changes in appearance of the growing body, the appearances of congenital abnormalities, and the recognition of normal variations. Many of the specific anatomical features of children were not appreciated before x-rays were discovered. This is demonstrated in the following examples.

A RETAINED BULLET

Charles Thurstan Holland was working as a general practitioner in Liverpool when on February 7, 1896, he saw some of the early x-ray work of Sir Oliver Lodge at Liverpool University College. Lodge took some radiographs of a boy who had shot himself in the hand. The wound had healed and it was impossible to tell if a bullet had been retained. A successful radiograph was obtained with an exposure of almost two hours, after several attempts had failed. Holland said that one cannot today imagine the excitement in the department when the plate was brought out into the daylight and the shadow of the bullet was demonstrated. By the end of May 1896, through the actions of his friend, the great orthopaedic surgeon Robert Jones, Holland had an x-ray kit. Robert Jones had immediately realised the value of x-rays in orthopaedic surgery.



SWALLOWED COINS

On October 2, 1896, Holland examined two small boys who had possibly swallowed coins. The examinations were not easy but coins were seen on a screen, and were removed the next day. One of the coins had been in the child's throat for over a year (Figure 1) and the child had a constant cough. The child had been treated for tuberculosis, and had been to many specialists and health resorts. Following the removal of the coin his cough disappeared, and Holland says how during this year of illness several hundred pounds had been spent on doctors and treatment, and this patient was one of his first great x-ray triumphs in diagnosis.

BRONCHIAL AND OESOPHA-GEAL FOREIGN BODIES

Holland became interested in foreign bodies in the airways, and saw that if they were opaque to x-rays they could be seen easily. One of Holland's cases was of an infant with an open safety pin in the oesophagus (Figure 2).

However, not all foreign bodies can be seen with x-rays. Peanuts could become stuck in the windpipe if inhaled and x-rays could be used to show a difference between lungs (indicating a stuck peanut without actually visualising it). This method is still common today, but was first reported in 1925 (Figure 3), and many lives have been saved this way.



Coin in oesophagus, October 2, 1896. Two-and-a-halfyear-old boy. Thirteen minute exposure, 6 inch coil, accumulator.



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An infant of 16 months with a peanut kernel in the right main bronchus. There is 'obstructive emphysema' of the right lung with increased transparency of the right lung, and marked shifting of the heart.

Open safety pin in the oesophagus.

SKELETAL DEVELOPMENT AND VARIATIONS

On September 1, 1896, Holland was able to examine a full term baby. He was fascinated to see the developing bones (Figure 4), particularly of the hands and feet. He realised the role that x-rays could play in anatomical studies and observing skeletal growth. He started collecting radiographs taken at different ages and in September 1896 showed them at the British Association in Liverpool (Figure 5).

This work on bone age was developed by John Poland from the Miller Hospital in Greenwich. Poland pointed out that the development of the bones differed quite considerably from that which had been previously described.

Information on normal child and skeletal development would be essential following the establishment of Well Baby Clinics, school health programmes, and the routine health examination of children in the first half of the 20th century.

In 1921, Prof. Thomas Wingate Todd in Cleveland, Ohio, began his studies of human growth and development. In 1931, three-month-old children were introduced into the programme and children up to the age of 14 years were entered into the study until the summer of 1941. Todd published his Atlas of Skeletal Maturation of the Hand in 1937. This groundbreaking book used data from the study group and also children from state-run schools and various social agencies. Todd found measurable differences between these two groups.

In 1950, William Walter Greulich and S. Idell Pyle, who were both anatomists from Stanford, published Radiographic Atlas of Skeletal Development of the Hand and Wrist, with a second edition appearing in 1959. This book remains the standard over 60 years later.

ANOMALIES AND VARIATIONS

On December 4, 1896, Holland demonstrated inherited abnormalities in a near-term baby. The majority of these congenital variations were unknown before x-rays were introduced, and it was largely due to the work of Alban Köhler of Wiesbaden that variations were first described.

Köhler was a founder member of the German Röntgen Society and became its president in 1912. The Lexikon der Grenzen des Normalen und der Anfänge des Pathologischen im Röntgenbilde was published by Köhler in 1910 and went through a number of German editions. The book was enormously influential and was illustrated using line drawings instead of x-ray images. It was translated into English in 1931 appearing as Röntgenology, the borderlands of the normal and early pathological in the Skiagram with a second edition appearing in 1935.

The work of Alban Köhler was continued by Theodore Keats from Charlottesville, Virginia. His Atlas of normal Roentgen variants that may simulate disease first appeared in 1973⁴ and is currently in its 9th edition. It is a modern classic and its presence in most, if not all radiology departments, is a witness to its value. As each new imaging technique develops, the normal and abnormal appearances need to be learned afresh.

THE EARLY YEARS

When considering the work of the pioneers I am impressed by the very high quality of the radiographs that were taken with such primitive apparatus. By 1900, excellent radiographs showing hip pathology in infants were being obtained. There was little understanding of the normal, yet alone the abnormal. There was no literature, no x-ray departments in any of the hospitals, and there were no experts. We owe them a huge debt.



A foetus at seven months. Radiographed on December 4, 1896. Five minute exposure, 6 inch coil, accumulator. The stillborn child had no nose, a deformed face, and six toes.

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FIGURE 5





The hand of a child aged one. Radiographed on September 17, 1896. Two minute exposure, 6 inch coil.

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WHY IMAGING CHILDREN I **IMAGING ADULTS**

BY FERMIN SAEZ AND PETER J. STROUSE

hildren are not small adults" is a classic mantra for all paediatric healthcare providers. Children are not adults from the point of view of anatomy (entirely different bodily proportions and development), physiology (body processes change with growth and maturation), and pathology (children and adults are affected by different disease processes and injuries). Imaging of children is therefore different from imaging of adults.

Some diseases seen in paediatrics are the same as in adults; however, they may manifest differently in the growing child. Many other disease processes are seen only in newborn babies, infants, or children and thus require different therapies and age-appropriate focused imaging approaches. Application of imaging protocols and paradigms used in adults is inappropriate for children more often than not.

diseased children of differing ages is fundamental for the proper choice of imaging modality, selection of proper imaging parameters, successful exam completion, and, finally, educated image interpretation.

CHOICE OF APPROPRIATE EXAMINATION

There is an array of possible imaging modalities for diagnosing paediatric diseases and injuries: plain x-rays (radiographs); fluoroscopic x-ray procedures (digestive tract imaging with the use of contrast media, a substance which is taken by the patient and enhances parts of the image); visualisation of joints with injection of contrast media (arthrography); angiography, as a guide for biopsy; ultrasound; Knowledge about the characteristics of healthy and inclear medicine; computed tomography (CT); and modalities are used in children.

A child's health and clinical presentation must be carefully considered before medical imaging exams are performed. When performed for a proper indication (i.e. a valid reason to undergo the examination), the clinical benefit of an x-ray imaging exam will outweigh the minimal risk associated with radiation exposure. However, efforts should be made to help minimise this risk by considering alternative imaging modalities that do not use ionising radiation or by using other non-imaging diagnostic methods.



magnetic resonance imaging (MRI). All of these

Children are more vulnerable to the effects of radiation due to the number of dividing cells and a long remaining life expectancy in which the side effects of radiation (possibility of developing cancer) could

manifest. Although these risks are very small, considering the greater vulnerability of children to the effects of radiation, the best protection is to completely avoid the use of ionising radiation by using radiation-free methods, namely ultrasound and MRI. With the exception of chest disorders, bone disorders and acute trauma, where x-rays and/or CT are usually done first, ultrasound is often the first choice of imaging modality for children and adolescents. MRI is often preferred to CT in children as a second line imaging modality.

Discussion between paediatricians, paediatric surgeons, orthopaedists (and other paediatric healthcare providers) and radiologists has become increasingly important in ensuring the most effective choice of imaging modality, given the continual advancement and new indications of modern imaging techniques.

AT THE TIME OF EXAMINATION

Performing an imaging study in a child is much different to performing the same study in an adult. It is the responsibility of the radiology department to use proper imaging protocols and parameters for the size of the child and for the indication for the examination. For instance, use of adult x-ray or CT parameters when imaging a child would result in excess radiation exposure without improvement of image quality. The number of images or type of images (positioning) may also be different compared to an adult examination.

Once the appropriate imaging modality is selected and a protocol is tailored to the child's age and clinical indications, there are still many challenges. Unlike adults, children cannot always understand the reason for their presence in a radiology department. They may be frightened and may be in pain and unable to cooperate due to crying and moving from the time they enter the room. Therefore, a child-friendly environment and dedicated staff are paramount for a successful examination. This presents a huge challenge for the radiographer, who must try to gain the child's trust and cooperation. Once cooperation has been achieved there is the challenge of keeping the child still throughout their imaging test. Some imaging tests are very quick – an individual x-ray takes a fraction of a second. Other imaging tests are lengthy – an average MRI examination takes 45 minutes to an hour. Ultrasound is relatively unaffected by motion as the images may simply be repeated, although a moving child makes ultrasound challenging as well.

The involvement of the child's parent or guardian is highly important to achieving successful imaging in many children. The presence of the parent and their comfort and instruction will often suffice to achieve good images.

Many dedicated paediatric radiology departments employ Child Life Specialists to assist children through imaging examinations. Child Life Specialists use distractive manoeuvres, play and comfort. They are particularly helpful for more involved or longer examinations such as fluoroscopic studies, CT or MRI.

For some examinations, sedation or general anaesthesia may be used, depending on the age of the child and their ability to cooperate, the type of examination and expected length, and whether there will be discomfort associated (i.e. an intravenous injection). Most commonly, this applies for MRI studies, which are longer in duration, and for image-guided interventional procedures (angiography, biopsies). "Knowledge about the characteristics of healthy and diseased children of differing ages is fundamental for the proper choice of imaging modality, selection of proper imaging parameters, successful exam completion, and, finally, educated image interpretation."

Fermin Saez / Peter J. Strouse

IMAGE INTERPRETATION

To diagnose a paediatric condition successfully, high quality images are needed. However, this is not the final step. The radiologist interpreting the examination must take into account the dynamics of a growing body, considering differences from preterm infants to large adolescents, and phases of organ and tissue growth and maturation. Thorough knowledge of paediatric physiology and pathology is mandatory in order to interpret the imaging results. In addition, effective review of the clinical history of the child is a key factor for correct interpretation of an imaging study.

At birth, the bones are only partially formed and are still partially composed of cartilage. In the long bones of the legs and arms and the short tubular bones of the fingers and toes, there are growth plates (the medical term is physis) at the ends of the bone. The bones grow at the growth plates, which are weak relative to more mature bone. Therefore, the presence of the growth plates predisposes the child to certain patterns of fracture not seen in adults (Figure 1). While most fractures involving the growth plates heal without complications, injury may result in premature fusion of the growth plates and loss of normal growth, resulting in unequal limb length. This complication does not occur in adults. The bones of a child are also different in composition to those of an adult - they are softer (medical term = plastic) than the bones of an adult. In children, fractures are often partial, buckling the surface of the bone or only extending part of the way across the shaft of the bone - patterns that are not seen with adult fractures.

The flat bones (i.e. skull, spine, pelvis, ribs) in an infant are also incompletely formed. At birth, the rigid portions of these bones are separated by cartilaginous portions, allowing for growth. For instance, the skull is composed of multiple component bones. At birth, these individual component bones of the skull are separated by cartilage and sutures, allowing for remodelling and growth with age. Growth of the skull does not cease until late adolescence, at which time the sutures fuse completely.

An example of a disease process unique to the paediatric musculoskeletal system is child abuse ('non-accidental trauma'). Fractures resulting from child abuse may have a specific appearance based on the mechanism of injury, the site of fracture and the lack of maturation of the bones (Figure 2). Findings may be florid (many fractures) or subtle (a single fracture mimicking the normal state). The diagnosis may be unsuspected. All radiologists interpreting radiographs of young children should be familiar with the imaging manifestations of child abuse.

A radiologist interpreting bone x-rays or head CTs of a child must be aware of the normal developmental anatomy of growth plates and the sutures of the skull. Without this knowledge, normal structures might be misinterpreted as a fracture or a fracture might be misinterpreted as normal structure (Figure 3). In addition, other non-traumatic processes may affect the growth plates and cranial sutures and the radiologist must be vigilant of any abnormality that might indicate an underlying disease process.

Nevertheless, many imaging examinations of children are performed by general radiologists whose practice mostly involves adult imaging. For example, in the United States, as many as 85% of paediatric CT examinations are performed at non-paediatric-focused facilities. A 2012 study looked at interpretation differences in images of children between general radiologists at non-paediatric facilities versus paediatric radiologists at tertiary care children's hospitals. For the general radiologists interpreting paediatric imaging, major disagreements in interpretation were found in nearly 22% of cases. The disagreement figures are much lower for



A 13-year-old boy with trauma to the right ankle. Anteroposterior (AP) ankle radiographs. A Salter I fracture is seen in the distal fibula (long arrow); the fracture courses through the growth plate (the width of the growth plate is abnormally increased, not involving the adjacent bone). This fracture has a good prognosis for healing without complication. A Salter IV fracture is seen in the distal tibia (short arrows); the fracture courses through across the growth plate involving the adjacent bone on either side of the growth plate. Salter IV fractures usually have a poorer prognosis which can interfere with future growth.

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FIGURE 2



AP radiograph of the right leg of a 3-month-old girl. Proximal and distal tibial 'bucket handle' fractures (arrows) can be seen. Such fractures are also called 'classic metaphyseal lesions'. This appearance is highly specific for the diagnosis of child abuse. This child's x-ray skeletal survey showed multiple other fractures.

dedicated paediatric radiologists: a study interpreting x-rays of four common injured joints (elbow, wrist, knee, ankle) found an error rate (including both relevant and non-relevant errors) of only 2.7%. Clearly, training and experience are of benefit to the proper interpretation of paediatric imaging studies.

Normal developmental findings may be confused with pathology (Figure 4). Common pathological processes, such as pneumonia, may be confused with more ominous pathology (Figure 5).

SUMMARY

The diagnostic imaging of children poses many challenges. Imaging of children is different to imaging of adults. These challenges must be addressed step by step, beginning with the choice of the most appropriate imaging test, performing the test appropriate for age and clinical indication, and properly interpreting the examination in a paediatric context.

Communication between the referring paediatrician and the radiologist is of great benefit in deciding whether to image, deciding on the best imaging modality, and for properly tailoring the examination to the patient. After an examination, communication back from the radiologist of unexpected or urgent findings hastens medical care. This 'before and after' collaboration saves time and expense, aids in minimising radiation exposure, and improves the comfort and efficiency for the patient and their family.



Five-month-old boy. Volume rendered image from a CT of the head. A right parietal fracture (arrowheads), which was initially mistaken for a normal variant on x-ray. This child was also the victim of child abuse. The other lines seen in the skull are normal sutures (arrows).



Eleven-year-old boy with painful knee after a fall. Lateral x-ray of the right knee shows irregularities/fragmentation of the posterior aspect of the lateral femoral condyle (arrows). This appearance is a normal developmental finding at this age, but inexperienced observers may confuse this with disease.

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Four-year-old boy with fever. Lateral x-ray of the chest. There are two opacities in the lung (pneumonia at the right upper and lower lobes). The lower opacity shows a rounded contour (circle), and could be mistaken for cancer. However, this appearance is relatively frequent for pneumonias in children ('round pneumonia'). If the patient were an adult, the finding would be more worrying and would be followed with a CT exam, whereas in a child, clinical treatment and a follow-up x-ray would be done.

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IMAGE GENTLY®: A BIG VOICE FOR OUR LITTLE ONES

BY DONALD P. FRUSH AND MARILYN J. GOSKE

-ray imaging saves lives. Medical imaging that uses x-rays includes radiography (such as a chest x-ray), fluoroscopy (an x-ray movie), and computed tomography (CT, sometimes called a CAT scan). These x-rays can be used to form pictures of the human body and provide valuable information that can help in the care of your child.

Why can x-rays 'see' inside the body? There are different types of radiation including everyday light, microwaves, and electromagnetic radiation from cell phones. To 'see' inside the body requires a type of radiation that is more energetic (so called ionising radiation). In general, the term 'radiation' usually refers to ionising radiation, such as the energy made in nuclear energy plants or in the past with the atomic bomb. We know that radiation at very high levels (high doses), from the atomic bomb for example, can cause harm and cause tissues to die. This is the reason for radiation treatment (or therapy) of cancers, like lung cancer in adults. High dose radiation can also cause harm

to healthy tissues, with effects such as skin burns, hair loss, cataracts (cloudy lenses) and the development of cancer. It is important to understand that the amount of ionising radiation used for medical x-ray imaging is very, very low compared with exposure that causes these kinds of damage. Doses used in medical imaging are usually hundreds to thousands of times lower than these tissue harming doses. We know there is little risk of tissue damage at these doses, but what the risk of cancer is at the lower doses used in medical imaging is not as clear. While we don't know if there is a risk (and if there is, it is very, very small) of cancer with the low doses of radiation used to take pictures in children, we must be very careful about protecting the children we image. For example, we have a responsibility to use these examinations only when necessary, and to use only as much radiation dose as is needed to provide helpful pictures.

Those of us who care for children understand that children are not small adults. They have different med-

ical problems compared to adults. A doctor would not think of giving the same dose of antibiotics to a football player as she would an infant. Similarly, for x-ray, doses must be based on patient size. Smaller children need less radiation dose than larger children to create appropriate pictures. Too much radiation should be avoided but too little radiation may not give detailed enough pictures.

Concern and lack of understanding about radiation used for medical imaging may come from patients and caregivers as well as the medical profession. It is vital to communicate clearly with all these groups. One method for providing information and guidance to parents and the public is through a technique called social marketing. Social marketing uses advertising or marketing techniques to reach a target audience to provide information about a topic for the purpose of improving behaviour to improve society. One organisation recognised the need for guidance about medical imaging. This professional organisation is the Alli-

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ance for Radiation Safety for Pediatric Imaging, known as the Image Gently® Alliance.

The Image Gently Alliance was formed in July 2007. The alliance leadership committee is made up of experts from four professional imaging organisations: the Society for Pediatric Radiology, the American College of Radiology (ACR), the American Association of Physicists in Medicine (AAPM) and the American Society of Radiologic Technologists (ASRT). These groups represent the critical triad of people taking care of children in medical imaging, the radiologists (physicians, with specialty training in medical imaging), radiological technologists (who perform the examinations), and medical physicists (responsible for the imaging equipment). The Image Gently Alliance leadership committee also includes media expertise as well as a patient advocate.

In addition to the four founding organisations, there are more than 95 affiliated professional organisations.

These include not only radiology and dental groups but also referring doctors (such as the American Academy of Pediatrics). In addition, the alliance has gone global, with over 25 international partners joining. Through these affiliations, the alliance represents almost one million medical and dental professionals.

The mission of the Image Gently Alliance is to promote safe and effective imaging care of children worldwide. How does the Image Gently Alliance work to fulfil its mission? The principal strategy of Image Gently is social marketing. Social marketing takes advantage of successful commercial marketing techniques through various media such as a website (imagegently.org), the internet, and information in both scientific journals and the public press, to promote the message of radiation safety for children.

The goal of the Image Gently campaign is to keep the message simple. The key message is: imaging saves lives but when you need to image, image gently, choosing the right test, and using the right dose (child-size). This message is positive and one of advocacy, rather than alarm. This alliance seeks to be independent and remain free from any true or perceived conflict of interest. For example, the alliance operations are not supported financially by manufacturers or other commercial interests. While the four founding organisations support the administrative costs, these expenses are kept to a minimum. The work of the alliance is largely performed by passionate volunteers who enable the organisation to operate efficiently and economically. The affiliate organisations do not make financial contributions, but do have a very important role in spreading the philosophy of Image Gently to its members.

What are the alliance's activities? A website has been developed to communicate with medical imaging professionals and the public. The website includes free, downloadable parent leaflets about various imaging procedures and has a frequently asked questions section. Educational modules and PowerPoint presentations have been prepared to help technologists better understand paediatric medical imaging, which have been translated into more than 15 different languages.

Over the eight years since the start of Image Gently, there have been six campaigns, each focused on one aspect of children's imaging. These include campaigns highlighting the benefits and risks of CT scans, interventional radiology (where x-ray studies help in



The message of the Image Gently Alliance is that for radiation use, "one size doesn't fit all"

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treatments), fluoroscopy, standard x-ray studies, nuclear medicine, and dental imaging. Three medical conferences have been hosted, and Image Gently representatives speak at conferences of other professional societies and organisations around the world. Dozens of articles about Image Gently have been written for both the public press and scientific iournals.

To date, Image Gently has achieved wide recognition and international acclaim for its efforts. Several publications have detailed the impact the content has made in changing radiology practice so that imaging is performed in a more child-friendly way, often with reduced radiation dose. The social marketing strategy, which raises awareness, provides education and promotes advocacy for paediatric radiation protection, has also been influential.

In the United States, a close relationship exists with Image Wisely[®], the adult counterpart started three years after Image Gently. Newer international medical radiation protection campaigns including EuroSafe Imaging and AfroSafe have also formed relationships with the Image Gently Alliance. In addition, Image Gently has worked together with the International Atomic Energy Agency and the World Health Organization on educational content about paediatric radiation protection.

There are challenges and opportunities as the Image Gently Alliance work continues to work at full speed. Volunteer efforts are essential, as committed professionals work with their heads, their hands and importantly their hearts. The needs are growing on an international scale and require firm partnerships to be sure that efforts are not being duplicated, and that they offer consistent and fact-based messages. It is easy to see that the extended relationships and cooperation achieved by Image Gently has been based on the shared need and recognition of appropriate childhood imaging, delivered in a positive manner and through modern and effective communication strategies to all those who are involved with this aspect of patient safety. There is an increasingly recognised need in many parts of the world and in many different aspects of imaging. And it is the right thing to do for children: one size cannot fit all.

www.imagegently.org



technologists

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RADIAIION ROTECTION IN **ROSAFE IMAGING**

BY GUY FRIJA AND PETER VOCK

ABOUT FUROSAFE IMAGING

As the demand for medical imaging examinations is constantly growing, safety and quality in radiological practice are more important than ever. The European Society of Radiology (ESR) took a major step in raising awareness of the importance of radiation protection in medicine with the launch of EuroSafe Imaging at the European Congress of Radiology (ECR) in March 2014. With its mission of supporting and strengthening medical radiation protection across Europe following a holistic, inclusive approach, the campaign provides a framework for the ESR's quality and safety initiatives.

EuroSafe Imaging is led by a steering committee that comprises representatives from ESR partner organ-

isations with a shared commitment to improving quality and safety for patients in Europe. Charged with setting the campaign's strategy and overseeing its implementation, the steering committee is chaired by former ESR President Prof. Guy Frija and consists of representatives from the ESR; the European Federation of Organisations for Medical Physics (EFOMP); the European Federation of Radiographer Societies (EFRS); the European Society of Paediatric Radiology (ESPR); the Cardiovascular and Interventional Radiological Society of Europe (CIRSE); the patient organisation European Federation of Neurological Associations (EFNA), on behalf of the ESR Patient Advisory Group; the industry group COCIR; and an observer from the European Commission.

campaign's aims:

- radiological procedures
- reference levels (DRLs)
- equipment
- and communication

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The steering committee's actions are guided by the

- To promote appropriateness and justification of
- To maintain radiation doses within diagnostic
- To promote the application of the 'as low as reasonably achievable' (ALARA) principle • To promote the use of up-to-date imaging
- To empower patients through better information
- To bring together a variety of stakeholders.

FUROSAFE IMAGING CALL FOR ACTION

EuroSafe Imaging's mission and objectives have been translated into a comprehensive strategy in the form of the EuroSafe Imaging Call for Action, which was launched in September 2014. The EuroSafe Imaging Call for Action is designed to support the International Atomic Energy Agency and World Health Organization's 2012 Bonn Call for Action, which identifies responsibilities and proposes priorities for stakeholders regarding radiation protection in medicine.











ACTION 8

Develop data collection project "Is your imaging EuroSafe?" and educational project on guidelines "Are you imaging appropriately?"



ACTION 9

Develop criteria for imaging procedures that use ionising radiation in specific exams and anatomical regions

·	J



ACTION 11

Improve information for and communication with patients regarding radiological procedures and related risks in order to ensure empowerment of patients



ACTION 12

Engage with other stakeholders and collaboration with related initiatives and regulatory authorities in Europe and beyond to contribute to a global safety culture in medical imaging

ACHIEVEMENTS

Since its launch in 2014, the ESR and its partners have successfully implemented several items of the Call for Action.

The development of the ESR's clinical audit tool. including the publication of 26 Level 1 audit templates, was completed in 2015. This year's European Congress of Radiology also saw the launch of ESR iGuide, a clinical decision support system for European imaging referral guidelines, developed by the ESR and National Decision Support Company (a US software firm) in partnership with the American College of Radiology. The ESR's new e-learning platform, Education on Demand, is now also up and running and contains eleven EuroSafe Imaging-themed radiation protection modules.

EuroSafe Imaging is also used as a framework to engage with European institutions, as the ESR has assumed the lead of a European Commission tender project on diagnostic reference levels for paediatric patients (PiDRL). In addition, the ESR and other medical associations collaborate with the research platform Multidisciplinary European Low-Dose Initiative (MELODI) and have developed a strategic research agenda for radiation protection that was open for public consultation until summer 2015.

EuroSafe Imaging also collaborates with stakeholders such as the Heads of the European Radiological protection Competent Authorities (HERCA), particularly regarding the process of justification for carrying out medical imaging procedures in the context of the Euratom basic safety standards directive. HERCA published a position paper on optimising dose in computed tomography (CT) in October 2014, according to which optimisation can only be successful if all involved stakeholders work together. The ESR supports this approach and strongly advocates a multi-stakeholder approach to radiation protection.

Moreover, a series of surveys entitled 'Is your Imaging EuroSafe?' has been launched as part of the ESR's EuroSafe Imaging campaign to improve radiation protection in Europe. The core aim of 'Is your Imaging EuroSafe?' is to build a European repository based on dose exposures for specific clinical indications that would be most helpful for self-benchmarking and for future establishment of diagnostic reference levels (DRLs). The surveys contain guestions on CT dose exposure values for different adult CT clinical indications. The preliminary results of the surveys on acute stroke and pulmonary embolus are already available online.

Another essential part of the campaign are its efforts to foster closer collaboration with patients through the ESR Patient Advisory Group, an arrangement that makes the ESR a role model for other medical organisations. The focus here is on providing patients with readily available and easy to understand information on medical imaging, and to encourage medical professionals to improve their direct communication with patients.

ROADMAP 2015-2016

On the basis of the 2014 Call for Action, The EuroSafe Imaging roadmap for 2015-2016 defines the priorities for the ESR and its partners for the current year.

Raising awareness of the importance of radiation protection is a central objective of the campaign, and to do so, effective communication is essential. The first priority of the roadmap is therefore to improve the EuroSafe Imaging website by making it more user-friendly, particularly for patients, and developing more informative and interactive content.

The second item on the roadmap is to develop guidelines for communicating with patients, which ties in closely with the expansion of the website content.

Advisory Group.

patients and carers

In addition, a new concept called 'EuroSafe Imaging Stars' will be launched this year, which aims at creating a network of key institutions across Europe with medical physicists and radiographers who are willing to support EuroSafe Imaging by providing information for data collection and benchmarking. In addition, qualitative interviews are envisaged to investigate key issues on medical radiation protection. The participating institutions will be named 'EuroSafe Imaging Stars', and acknowledged on the EuroSafe Imaging website and in media and press activities.

The ESR is also in the process of planning a project on dose management and developing proposals for the International Commission on Radiological Protection (ICRP) to consider the establishment of diagnostic reference levels (DRLs) for the most frequently occurring clinical indications, as well

Paediatric Imaging

As a 2013 ESR survey on patient communication shows, there is significant room for improvement, as only one third of radiologists receive training on communicating with patients. The aim of providing guidelines on this subject is to support radiologists with aspects of daily practice, such as giving bad news to patients or explaining the benefits and risks of a specific imaging procedure. This will be done in close collaboration with the ESR Patient

Radiation protection in paediatric imaging is an issue of particular concern for EuroSafe Imaging, which is why including paediatric patients front and centre in the campaign is a further key aspect of the annual action plan. In cooperation with the European Society of Paediatric Radiology, the ESR is exploring ways to initiate new projects in this area based on the European Commission's recommendations that will follow the completion of the PiDRL tender project. The objective of these efforts is to use EuroSafe Imaging as a platform for professionals as well as as those examinations that require the highest levels of exposure.

INTERNATIONAL OUTLOOK

EuroSafe Imaging also looks to have an impact beyond Europe's borders, which is why the ESR is engaging with related campaigns such as Image Wisely and Image Gently in the United States. By fostering cooperation and a regular exchange of information, synergies are created and all partners can learn from each other's efforts in putting radiation protection at the heart of daily practice.

Beyond working with other campaigns, EuroSafe Imaging also aims to be a role model for regions where no prominent radiation protection initiatives exist. The ESR has therefore proudly supported the launch of AFROSAFE at the 2015 Pan-African Congress of Radiology in Kenya in February 2015. In Canada, radiologists initiated a coalition in 2015 called Canada Safe Imaging to promote radiation protection.

Achieving progress internationally necessarily involves engaging with international organisations. Through EuroSafe Imaging, the ESR seeks to contribute to keeping radiation protection on the World Health Organization's (WHO) and the International Atomic Energy Agency's (IAEA) agenda, as outlined in the Bonn Call for Action, and uses every opportunity to get its message heard. In cooperation with the International Society of Radiology (ISR), radiologists had a proposal accepted to hold a side event on paediatric imaging during the WHO's 2015 World Health Assembly, held in Geneva in May, during which EuroSafe Imaging Steering Committee Chair Prof. Guy Frjia presented the campaign's activities focusing on children. EuroSafe Imaging has also been presented during several major international radiology congresses, in Japan, Brazil and Turkey, among others.



Gemeinsam für Patientensicherheit

FRIENDS OF EUROSAFE IMAGING

The concept of Friends of EuroSafe Imaging aims to demonstrate that the ESR's drive to strengthen radiation protection is supported by a great number of radiologists, other medical professionals, patients and members of the public. We would also like to encourage you to support us in our mission to improve quality and safety for patients in Europe and beyond by signing up as a Friend of EuroSafe Imaging at

www.eurosafeimaging.org

Ensemble pour

pour la sécurité du patient

יחדיו למען

Razem dla bezpieczeństwa pacjentów

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HE IMPORTANCE

BY JENNIFER GREHAN

Here we will talk about the importance of a child-friendly imaging environment to patients, staff and parents/carers. Let's start at the beginning and take a 'walk' through things we might consider.



MMOBILISATION

ROOMS (RECEPTION, WAITING AND IMAGING)

EAM AND TRAINING

A VAILABILITY

NFORMATION

D ISTRACTION

E QUIPMENT

A TMOSPHERE

ARENTS, PREPARATION AND PLAY SPECIALISTS



Both adults and children usually fear the unknown. For a child, diagnostic imaging is a world of equipment, strangers and shiny technology and, as a service provider, most departments are always bustling and busy. We can't change the purpose of hospitals or imaging departments, but we can make these environments as friendly as possible for children.

PREPARATION, PARENTS AND INFORMATION

Apart from emergency cases, childhood imaging will often be by an appointment system, allowing for a degree of preparation. By preparing both the child and parent/carer with information delivered in advance, the examination itself comes as less of a surprise. Age-appropriate patient information can take many formats, including printed information sheets, booklets included with appointment letters, websites where examinations are explained through text, pictures and podcasts, and downloadable apps for mobile and tablet use which take the patient and parent/carer on an interactive journey through what to expect during the examination.

"There is no knowledge that is not power" - there is a lot to be said for giving some control back to a parent/carer who may feel out of control with regard to what is happening to their child. In introducing a child and their parent/carer to information early, we include them in patient-centred care and have the potential to turn a degree of anxiety into anticipation. Involving parents or carers in why the child needs to stay still, and the use of lead protection, helps them to anticipate the steps in an imaging examination. Parental involvement in encouraging the child's participation usually increases the chance of success.

ROOMS, ATMOSPHERE AND DISTRACTION

Patients usually arrive at the reception desk. From the moment of arrival it is very important the child feels included and important. Reception desks can be multiheight (Figure 1) to allow for immediate eye contact with smaller children or those in buggies as well as any wheelchair-bound population. Wipeable art at eye height is a great distraction in any open area (Figure 2).

Children are familiar with play. At home, in nurseries, at school, it is a very normal part of their world. Opportunities to play in the waiting area promote a relaxed atmosphere, and distraction is a useful technique for the parent and child. Careful planning of even the smallest of waiting spaces can yield rewards with the use of wall space for bright murals, wall mounted screens for DVDs and racks for interactive books. With larger spaces, waiting areas for different ages can be considered: seating for parents/carers, a safer area for smaller babies, a messy play/art area for younger patients (Figure 3) and a separate area for the adolescent population.

Making imaging rooms child-friendly is more challenging, as they will have at least one large piece of equipment dominating the room. The important thing to remember here is visual engagement – there is very little that we can do about the size, shape or location of the equipment, but what can we control? If walls





area space

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Multi-height child-friendly reception desk

GURE 2



Wipeable eye-height artwork along an open space

Messy play/artwork area within a larger waiting

need to be a standard neutral colour, white or cream, cupboard doors can be a brighter colour – yellow or red. This immediately lifts the focus and distracts from equipment. In paediatric imaging, a theme often helps staff tell a story which again encourages familiarity and acts as a distraction: 'Welcome to the Circus' – lions/tigers/elephants; 'Under the Sea' – fish/mermaids; 'Outer Space' – spaceships/astronauts/aliens are all examples of themes that can be integrated into the imaging setting.

Infection control needs to be foremost in the mind of anyone making a space child-friendly. Cuddly toys are not usually easily cleanable, but wipeable, colourful mobiles to hang from ceilings work well, as do light projectors and displays that can be moved between rooms. When there are issues with murals being painted onto walls, wipeable life-size stickers can be sourced as a flexible replacement. Where cost is an issue, blackout roller blinds can be attached to doors and painted with a colourful animal or character in support of the theme in the room. Large pieces of scanning equipment such as CT or MRI can be painted with entertaining colours or themes. The bright cupboards and drawers should house all ancillary and immobilisation equipment leaving surfaces free for toys and easy access for cleaning. In-room toys need to be wipeable, durable and wherever possible, noisy!

PLAY SPECIALISTS AND SEDATION/GENERAL ANAESTHETIC

Some imaging tests take longer than others and movement during an imaging examination can be detrimental, either making the image more difficult to interpret correctly, or making it necessary to repeat parts of the examination. Other techniques may be required to keep a child still during the imaging exam. Sedation is the administration of medication to a patient specifically with the purpose of making them sleepy enough that they may just lie still and un-distressed for the duration of the scan. General anaesthetics are stronger and are used to put a patient completely to sleep and control the moment at which they should wake up.

Hospitals dealing with children will often have access to play specialists. These professionals are experts in using play as a way for children to interpret the hospital world around them and understand what is happening to them. Some imaging departments use the play specialist team to work with patients and parents/carers to improve the overall imaging experience and in some cases (alongside faster scan times) this decreases the need for sedation or general anaesthetic.

EQUIPMENT, IMMOBILISATION AND AVAILABILITY

Equipment in a child-friendly imaging department can mean many things. We have covered imaging equipment in terms of visual appearance, but it also needs to be user friendly. What use is a lovely, colourful fluoroscopy suite that is so loud when it moves that it sounds like a rocket being launched? Or a bright and shiny diagnostic x-ray unit which takes one whole minute to change the settings? Paediatric imaging requires thought and foresight, and after that, the equipment needs to be adaptable so that changes can be made quickly and easily.

It is important not to forget the need for ancillary equipment to be child-friendly. Lead rubber aprons

FIGURE 4



Staff wearing col aprons and suits

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Staff wearing colourful, distracting lead rubber



Parent holding patient using a variable height chest stand and Velcro strap to help with immobilisation

or suits should be available in a wide range of sizes and, where paediatric patients are concerned, brightly coloured patterns (Figure 4). Used correctly, these can be as much a distraction tool for a young patient as a loud rainmaker or a musical book. Like lead rubber aprons, the most important thing with other equipment such as foam pads, gonad shielding, sandbags or Perspex blocks is that they should be in a variety of sizes and in plentiful and local supply. Availability and supply is key.

As for the necessity that is immobilisation, it is not about fancy devices and tools; it is about parents/carers and, wherever possible, familiarity. Examples might be blankets for gentle swaddling or Velcro leg straps which can be likened to a seat belt in a car seat (Figure 5).

SPACE, TEAM, TRAINING AND COMMUNICATION

As with all aspects of healthcare, the room should operate smoothly around the patient: from parent/ carer to radiographer to radiologist. Communication is the key to successful work in the best interests of the patient. An appropriately planned imaging environment helps encourage this. When we think of children we naturally think smaller, but children often require more space than adults, rather than less. This is partly due to the fact that children rarely present to imaging departments unaccompanied, and space plays a huge part in the ability to accommodate their parents or carers. Another factor is that, in as many cases as possible, we should be looking to adapt a technique to each individual child, which in itself requires room to manoeuvre.

A responsive, confident, collaborative team is vital, in order to encourage as much child-friendliness as possible. This is doubly important in paediatric imaging where time, accuracy and speed are such decisive factors. In this respect, appropriate paediatric training and insight of staff is extremely important, because children need to be treated very differently to their adult counterparts, both from an engagement and imaging perspective.

CONCLUSION

The importance of child-friendly imaging facilities is paramount. A well-planned environment has a major influence on both the quality of the examinations and the overall experience of a child and their family.

"For every minute spent organising, an hour is earned" Benjamin Franklin

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Jennifer Grehan



GLOBAL PAEDIATRIC RADIOLOGY

BY M. INES BOECHAT, RUTGER A.J. NIEVELSTEIN, VERONICA DONOGHUE

s the specialty of paediatric radiology grew, professional societies started to form around the world. The North American Society for Pediatric Radiology (SPR) was founded in 1958 and the European Society of Paediatric Radiology (ESPR) in 1963. These were followed more recently by the Latin American Society of Pediatric Radiology (SLARP) and the Asian and Oceanic Society for Paediatric Radiology (AOSPR).

The organisations have been very active and have made significant contributions to better imaging care of children. The respective leadership of these societies held several meetings in the mid-2000s to discuss the possibility of better coordination of their efforts and unified representation in international forums, such as the World Health Organization. Thus, in 2011, the World Federation of Pediatric Imaging (WFPI) was founded during the International Pediatric Radiology meeting held in London. The purpose of the federation is to provide an international platform for the united paediatric radiology organisations to address the challenges in global paediatric imaging training and the delivery of services (Figure 1). The founding president of the WFPI, Prof. M. Ines Boechat, was assisted by an exceptional group of leaders from the four existing international societies, in creating the new organisation's bylaws, mission statement, Council of Directors and Executive Committee, as well as the permanent committees on Education and Outreach. Efficient managerial, strategic and creative support was provided by Ms. Amanda Dehaye, based in France, who helped the federation to move forward rapidly. By August 2012, the WFPI strategic vision was in place. Its main goals are:

- Communication and collaboration between paediatric imaging practitioners, via their organisations
- Advocacy of appropriate practices and resource allocation
- Education
- Patient safety, in particular radiation safety and protection
- Outreach and training in lower resource settings
- Research
- Information
- Institutional high performance

It soon became clear that the whole African continent was lacking representation. African paediatric radiologists, although small in numbers, were full of enthusiasm; by 2013, the South African Society of Paediatric Imaging (SASPI), and the Society of Pediatric Imaging in Nigeria (SPIN) had been formed under the umbrella of the African Society of Paediatric Imaging (AfSPI). It was with great joy that this organisation joined the founding members at governance level.

Other national and international organisations have joined the WFPI over the years - the British Society of Paediatric Radiology (BSPR), the German Paediatric Radiology Society (GPR), the Spanish Society of Pediatric Radiology (SERPE), the French Society of Pediatric Imaging (SFIPP), the Indian Society of Paediatric Radiology (ISPR), and the Argentinian Federation of Diagnostic Radiology and Radiation Therapy Associations (FAARDIT). As its sphere of influence increases, the WFPI expects that more organisations will join.

Because the WFPI is an organisation with membership spread across the entire spectrum of time zones,



the issue of communication is crucial and a strong website with many tools was a fundamental requisite.

The initial site, www.wfpiweb.org, created with the support of the ESPR and SPR, has grown to a very solid and versatile structure. Social network tools have expanded, with Facebook, Twitter, and YouTube accounts recruiting new followers on a daily basis. You are invited to explore the different areas of the website, which has a strong educational component, funded by the SPR and ESPR.

Outreach goes hand in hand with education; since its inception, the WFPI has explored different ways to support global health efforts led by its members. Several scientific articles, illustrating the WFPI outreach efforts were published as a special issue of the *Pediatric Radiology* journal in June 2014¹.

As Africa, Asia and Latin America have the largest numbers of children and significant disparities in access to healthcare, the focus of our work has centred there, with telemedicine projects in collaboration with the American College of Radiology, Médecins

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Sans Frontières, Image the World and RADAID, among others. A worldwide network of volunteer paediatric radiologists is in place to participate in projects originating in diverse locations, such as South Africa, Mozambique, India, Cambodia and Laos.

Work on radiation safety, a topic of particular importance in children, has been done in collaboration with the Alliance for Radiation Safety in Pediatric Imaging, a coalition of healthcare organisations dedicated to providing safe, high quality paediatric imaging worldwide. Its ultimate goal is to change practice patterns via the Image Gently and EuroSafe Imaging campaigns. European and Latin American paediatric radiologists

have also actively participated in meetings held by the World Health Organization (WHO) and International Atomic Energy Agency (IAEA), contributing to the development of important documents, such as the Bonn Call for Action in 2012². New developments include the creation of the AfroSafe campaign and discussions to establish a similar LatinoSafe initiative.

Much has been achieved since the WFPI was created; as a new organisation, it faces many challenges. However, with the support of its members, we all hope that it will continue to grow and became a stronger voice advocating better imaging care of children around the world.

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WFPI Logo, developed by Ms. Francisca Soto, from Chile

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PAEDIATRIC IMAGING TECHNIQUES

- RADIOGRAPHY
 - RASOUND
 - 1PUTED TOMOGRAPHY
 - INETIC RESONANCE IMAGING
 - RID IMAGING
- INTERVENTIONAL RADIOLOGY
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RADIOGRAPHY

BY BELLA SAID, CATHERINE OWENS AND OWEN ARTHURS

hildhood, from birth to 18 years of age, is a time of rapid development and growth on a physical, psychological and emotional level. Radiology plays a vital diagnostic role within paediatric healthcare, and millions of young patients undergo imaging examinations using x-rays all around the world (UNSCEAR, 2008). This chapter will give an overview of the important role that imaging plays, and how a team of radiology nurses, play specialists, radiographers, sonographers and radiologists work together to administer the best possible patient-centred care. Paediatric radiology encompasses diagnostic imaging, treatments such as radiotherapy, and interventional procedures, which all take a minimally invasive approach to the patient care pathway.

The commonest imaging investigation in children is the normal standard x-ray. The term 'x-ray' is used to describe both the procedure taking place, and the image that is created. An x-ray image is acquired by transmitting x-ray particles (radiation), from an x-ray tube, through the patient to create an image on the other side (the image receptor). Some structures block x-rays due to their density and therefore appear white in the resulting image (e.g. bone; Figure 1), while less dense structures (e.g. air) allow radiation to travel through, and so appear black (e.g. Figure 2). Various shades of grey in between depend on the density of the tissue. Radiologists also add materials such as iodine, a contrast agent, often given by mouth or via a vein, which also block x-rays and so make certain structures in the body more visible on the x-ray image.

We use these simple black and white images to assess several important features of growth, development and disease. For example, x-rays of the hand can allow us to assess skeletal maturity (Figure 1), while x-rays of the spine can tell us about abnormalities as well as their successful treatment (Figure 2). Other conditions are more complex in nature, such as genetic or inherited conditions. Cystic fibrosis, for example, can be imaged using conventional chest x-rays (Figure 3) before needing more complex imaging.

X-rays have developed over the years and moved away from the original x-ray film used to create an image. This contained thousands of miniscule crystals that on impact with x-rays enable us to get the well-

FIGURE 1



Hand x-rays showing how the bones mature with age. A one-year-old child's hand (left) compared to a 16-year-old's (right) which has almost stopped growing. Hand x-rays such as these are useful to estimate how much more growth to expect.



known black and white x-ray image. At the time, the film had to be developed with various chemicals and then dried. This film has been replaced with computerised detectors that eliminate the need for chemical processing, speeding up the process. Images can now be altered with computer software to show different body parts, whereas originally, more than one x-ray exposure would have been required.

Although quick and easy to acquire, which is why they are so commonly used, the actual rays are only present for less than a second at a time and are not used continuously. As a result, x-ray images are taken like photographs, a snapshot in time. This is why staying still is so important, otherwise the images will be blurry, like trying to photograph someone running past. Children often need help to stay still, such as a foam pad to rest against, a seatbelt to hold the child in position, or simply the parent's hands helping and comforting a child in the right position.

HELPING CHILDREN UNDER-STAND X-RAYS

When a patient of any age comes in for an x-ray for the first time, the unfamiliarity of the situation generates anxiety. Paediatric radiographers are not only specialists in using the equipment and understanding the technological factors behind getting a good image, but also in the human aspects of helping a child in an unfamiliar situation.

Every radiology service has to ensure that patients and families are given all of the information necessary to make an informed decision. Within paediatric radiology departments this will vary greatly, as the patient could be a baby or an 18-year-old, so this means that health professionals need to assess and recognise the needs of the patient and information needs to vary accordingly. The need of a patient also ing relationship is rapidly built up between the family

varies according to gender, race or ethnicity, religion or belief, and disability (Lansdown, 2011). While babies are unable to be involved in decision making, most young children enjoy being involved in the process. This is achieved initially by explaining the unfamiliar room, which enables children to engage with the new environment. As well as allowing children to familiarise themselves with the room, there are a variety of things one can do to help, for example, by moving the x-ray table up and down or using toys. Familiarity helps to overcome the initial anxiety which children may experience (Chesson et al., 2002).

GETTING GOOD IMAGES IN CHILDREN

The younger the patient, the more creative the radiology team needs to be and creativity need not be expensive. A few sheets of paper and coloured pencils have been proven to help children express their anxiety and fear (Chesson et al., 2002). Equally, music has been found to be a good distractor. Parents or health professionals may sing to or with the children or music can be played on mobile phones. Any distraction techniques that are used at home can work just as well in a hospital environment. Sometimes a pre-prepared flip-book of what is likely to happen can help, or explaining the situation using the child's toys to mimic what the child needs to do. Parents and carers can help to explain that they will stay close to the child, as well as what the machine might sound like. An x-ray image of a toy can help to explain the purpose of the examination, serving as both a visual aid to bridge the gap in the child's understanding. Often the first few minutes of the interaction are crucial for the success of the examination, and everyone who works with children will recognise the importance of a smile and open body language. For the brief time the child may be in the imaging department, a trust-

FIGURE 2



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X-rays are used in assessing the spine for curvature. Here, a 15-year-old has undergone surgery (right) to correct a curvature of the spine (scoliosis; left).

and the radiology team, working together to make the experience exciting and easy for the child.

For children with learning disabilities or language disabilities the experience can be even more challenging and often a longer period of time is required to familiarise patients with the imaging environment. Children are encouraged to become part of the learning process: e.g. learning that an x-ray is required to check for the progress on his or her disease. The explanation provided by the radiology team helps to develop the patient's understanding of the matter, and adults can support and nurture the idea that they are involved in their own healthcare.

One of the vital skills involved is the ability to act and react to a situation. Radiographers provide an explanation that is age appropriate and wait to see if it has the desired effect. The radiographer then has to react and see if further explanation is needed. This interaction is a three-way street between the radiographer, the patient and the family accompanying them. It is a relationship based on respect and built on the passion of professionals who work to provide a patient-centred approach. This kind of relationship, where the health professional and parents/carers inform the child or young person about what is happening, empowers the child to feel able to express their concerns, and makes sure that they are listened to. However, in some difficult situations, not all young people want to be informed and involved in their decision-making, and

the experience of the imaging team will help to tailor the process to the individual child.

CHILDREN AND RADIATION

The easiest way to minimise the radiation dose received by children during x-ray imaging is to only image the body part required, and by blocking off areas which are not important, using lead shielding. Radiographers are required by law to 'justify' each x-ray exposure (to ensure that it is medically necessary) and to carry out the correct examination in the best possible way. A lot of research has gone into improving x-ray examinations throughout the years and one of the central aims of all radiology departments is to minimise the radiation dose used in obtaining these types of images. Other dose-minimising techniques are also used in order to get the best possible image while using the smallest amount of radiation possible. A chest x-ray examination now involves as little radiation as a few days of normal background radiation. Other body parts (such as the spine; Figure 2) may require higher doses, because a larger body part and more bones are being imaged, but the lowest dose possible is still used.

Paediatric radiology teams work closely with the patient and their family to ensure that the best possible image is obtained with the least inconvenience and within the shortest space of time, to help the child's diagnosis and management.

FIGURE 3



Chest x-rays can be used to demonstrate severe lung disease such as cystic fibrosis in this 13-year-old girl. The abnormal lung areas stop the x-rays penetrating through and appear white (left). The same patient following lung transplantation (right).

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ULTRASOUND

BY BRIAN COLEY

fter x-ray imaging, ultrasound is the most versatile and valuable imaging method for children. Unlike most techniques, ultrasound uses no radiation to produce images of the body, which is especially important when taking care of children. Modern equipment allows detailed imaging of many parts of the body, the evaluation of blood flow, the determination of tissue stiffness, and with ultrasound contrast agents the quantification of tissue perfusion. Ultrasound is also used as a treatment method to destroy tumours and to facilitate the delivery of medication and gene therapies.

DIAGNOSTIC ULTRASOUND

How does ultrasound work?

As the name implies, ultrasound creates images by sending high frequency sound waves (above the level of human hearing) into the body and then listening to the echoes of those sound waves after they have been reflected by internal structures. This happens through placing a transducer (also called a probe) on the skin surface with the help of a coupling gel. To cre-

ate images, there needs to be an 'acoustic window' so that sound can enter the body. One of the limitations of ultrasound is that bone and air block the transmission of sound meaning that images cannot be created beyond them. Sound waves are attenuated logarithmically as they travel through the body, so imaging very large people is more difficult. When sound waves return to the probe, they are converted into electrical signals that computers in the ultrasound machine process to create an image of the structures inside the body. The physician or technologist moves the transducer to acquire images from different angles to create a complete picture of an area of interest. Depending on the medical question that needs to be answered, a number of different ultrasound techniques may be used:

B mode, grey scale, or conventional ultrasound imaging

This is the most commonly performed ultrasound examination, and it allows depiction of internal anatomy and tissues. Sound does not travel the same way through different tissue types, which allows the distinction of different parts of organs, the differentiation of fluid from solid, and the detection of calcifications and stones. Extended field of view imaging allows the piecing together of many individual images to create more extensive images and relationships of anatomy (Figure 1). Newer transducers allow the acquisition of whole volumes of information that can then be used to create multiplanar and even three-dimensional displays of structures.

Evaluation of the kidneys and bladder is one of the most common paediatric ultrasound examinations. Ultrasound readily depicts dilatation of the collecting system that may indicate obstruction or vesicoureteral reflux, scarring from previous injury, and congenital abnormalities and malformations. The evaluation of abdominal pain and masses is also very common, and ultrasound allows detailed examination of the liver, gall bladder, pancreas, kidneys, and other organs. In a young child with an open fontanel (the 'soft spot' on the head), ultrasound allows high resolution imaging of the brain (Figure 2) to assess for malformations, bleeding, and hydrocephalus (water on the brain). More recently, ultrasound has been used to evaluate chest diseases such as pneumonia and tuberculosis.



M mode imaging

M mode imaging depicts the movement of internal structures over time. This method is most commonly used in imaging the heart (echocardiography) where it can help to quantitate the movement of heart valves and the amount of contraction of the cardiac chambers. Other uses include evaluation of movement of the diaphragm, the spinal cord, and ureteral peristalsis.

Doppler ultrasound

By looking at differences between the sound frequencies that the transducer sends out and the frequencies of the sounds that return, one can determine if the sound has bounced off something that is moving (like blood in an artery or vein). The Doppler effect allows quantification of this motion, so that we can determine how fast blood in a vessel is flowing. Pulsed Doppler creates an image of this velocity over time and can help determine things such as whether there is an abnormal vessel stenosis (narrowing) or if there is a problem with blood flow reaching an organ. Colour Doppler uses the same techniques, but depicts flow as moving colour on the grey scale image. This is extremely useful for making sure that blood vessels





Extended-field-of-view longitudinal sonogram of a newborn's spine shows the entire spine from the tip of the sacrum (arrow) to the last thoracic vertebral body (T12). The end of the spinal cord (*) is in normal position.

FIGURE 2



Midline sagittal sonogram of the brain in a term infant shows a normal corpus callosum (arrowheads), normal 4th ventricle (arrow), and a normal cerebellum (C).



inflammation.



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Sonogram of the right lower quadrant in a nineyear-old boy with pain and acute appendicitis shows an enlarged appendix (A) with increased blood flow to the wall (orange color) indicating



Shear wave elastogram image from infant with cholestasis. Shear wave speed measurements are markedly elevated (indicating abnormal stiffness), and the liver is extremely heterogeneous. Biopsy performed the same day confirmed cirrhosis due to biliary atresia.

(Image courtesy of Jonathan R. Dillman, MD)

Voiding cystosonography examination in a young child detecting ultrasound contrast agent (orange) in the right renal pelvis indicating vesicoureteral reflux.

(Image courtesy of Kassa Darge, MD)

are open and that flow is in the proper direction, and for indicating whether more detailed examination with pulsed Doppler might be necessary. Increased colour Doppler flow can also indicate abnormal inflammation (Figure 3).

Elastography

Elastography is a newer technique that allows ultrasound to determine how hard or soft a tissue is. There are different types of elastography, but they all are based on the fact that sound travels faster through stiffer tissue than it does through softer tissue. By measuring with special ultrasound pulses, these differences can be determined. Some methods display the differences in tissue stiffness as different colours, whereas some allow quantification of differences reported as changes in the speed of sound or in the pressures created by the sound waves (Figure 4).

Elastography has been used to help distinguish between benign and malignant masses, as cancers tend to be harder and stiffer than benign tumours and normal tissues. This has shown use in many adult diseases, and shows promise in paediatrics in evaluating fibrosis, tumours, inflammation, and other conditions.

Contrast-enhanced ultrasound

Ultrasound works by sound interacting with tissue to create an echo that is received by the ultrasound transducer. Sometimes these echoes are very weak and the resulting image is poor. Ultrasound contrast agents are gas-filled microbubbles that resonate when struck by an ultrasound beam. By evaluating the returning signals that are specifically from the contrast microbubbles, more sensitive and detailed images can be obtained. Despite a very good record of safety, however, these agents are not approved for use in all countries (such as the United States).

When administered through a bladder catheter, ultrasound contrast agents enable the evaluation of vesicoureteral reflux (where urine travels backwards from the bladder to the kidneys). This technique (voiding cystosonography) is as sensitive and specific as traditional imaging methods but without the need to expose the child to radiation (Figure 5). When given intravenously, ultrasound contrast agents allow very sensitive depiction of blood flow and the quantitative determination of tissue perfusion. Different perfusion patterns can help distinguish benign from malignant liver masses thus removing the need for further imaging and even biopsy. By increasing sensitivity to blood flow, ultrasound contrast agents are also proving useful in the evaluation of inflammatory bowel disease and juvenile idiopathic arthritis.

THERAPEUTIC ULTRASOUND

Ultrasound is most commonly used to make diagnoses. High-intensity focused ultrasound (HIFU), however, is a promising use of ultrasound as a therapy. All ultrasound causes some heating of tissues. The power outputs of diagnostic ultrasound machines are limited to a level that minimises the heat created so that no harm is done. HIFU systems have a much greater power output, and their specialised transducers focus the ultrasound beam like light is focused with a mag-

with gene therapy.

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nifying glass. This allows the generation of very high temperatures that can be precisely directed within the body. When used in conjunction with MRI guidance and temperature monitoring, very precise targeting and heating of tissues can be performed.

The primary use of HIFU has been to destroy tumours within soft tissues and bone without requiring an incision. Tumours of the liver, kidneys, prostate, uterus, and extremities, and primary bone tumours and metastases have been successfully treated, alleviating pain and avoiding surgery. Specialised HIFU techniques are being developed that are helping to increase the effectiveness of chemotherapy and to improve results

CONCLUSION

Ultrasound's versatility and avoidance of radiation make it an ideal imaging tool for examining children. When performed by a paediatric specialist with proper training and experience, ultrasound is often the only imaging method needed to arrive at a correct diagnosis. Continued development of newer diagnostic and therapeutic methods will only increase ultrasound's value in paediatric healthcare.

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TOMOGRAPHY

BY RONALD BOOIJ AND NANKO DE GRAAF

he purpose of imaging is to look inside the human body and to find answers for complaints or unclear situations. Imaging can be done by several methods, for instance with magnetic resonance imaging (MRI) which uses a very strong magnetic field, ultrasound (as for instance used in imaging unborn babies), or computed tomography (CT). This chapter will focus on the use of CT in children.

A CT scanner is a doughnut shaped machine with a table and uses x-ray radiation. The x-rays are generated in a box, called the x-ray tube, and sent through the body while rotating around it. The x-rays are measured by the machine with a detector system on the opposite side. This system measures the intensity of the radiation passing through the body and thereby calculates how much the x-rays have been attenuated (reduced) on their way through. In this way information from different angles of the body is received and allows the computer to create images of the body. These cross-sectional images made with the CT scanner are called slices. Normally the patient will lie on

their back on the CT table while cross-sections of the body are made. With computer techniques it is possible to arrange these slices in such a way that you can view images of the body from different points of view. For instance, to arrange the images in a way that shows the patient from the front, or back. CT may show very detailed information of any structure inside the body with highly accurate diagnostic sharpness. For instance, fine details of bone structures in the limbs or the bony structures inside the ear, complex birth defects and diseases. Not only two-dimensional images but also three-dimensional images can be created. Colours can be added to highlight specific details as illustrated in Figure 1.

CT technology is widely available; practically all hospitals in the western world have at least one CT scanner. CT was invented in the early seventies and consisted of one x-ray tube and one detector, which made the time taken for the examination very long (several minutes and several hours to reconstruct). At the end of the eighties, examination speed was increased with the

patient or parents.

Imaging of children is one of the most challenging and exciting fields in CT imaging: they are small, difficult (or impossible) to instruct and often they don't want to lie still. The cooperative child can be positioned on the table with the arms down when imaging the head and neck. The arms have to be put alongside the head when imaging the lungs, heart, abdomen and legs. This is because the bones in the arms can cause visual artefacts in the images, which can lead to misinterpretation or even 'hide' important structures.

When it is difficult to persuade the child to lie on their back on the CT table, it is sometimes helpful to use a special mattress: a vacuum cushion, a bag filled with foam balls. After the child is wrapped in the cushion, most of the air in the cushion is sucked out, creating a firm mattress. This will prevent unwanted movement, without harming the child. Figure 2 shows a doll representing a child positioned for CT in a vacuum cushion.



introduction of new scan possibilities which allowed movement of the table during the image acquisition process, the so called multi-slice spiral CT.

CT images are created with the same principles as in normal photography: voluntary or involuntary motion makes it difficult to create clear images. Ongoing technological improvements mean that most recent CT scanners are quite fast and where general anaesthesia was often needed in the past for the patient to lie completely still during a CT scan, this is often no longer required. It is nowadays even possible to image the paediatric beating heart due to the presence of more detectors, increased rotation and acquisition speed and even the use of two x-ray tubes and detectors. The heart itself can be imaged with special scan techniques using the electric signals (ECG) of the heart, fast imaging and appropriate instruction of the



A 3D image of a 2-year old child showing the large blood vessels to and from the heart, highlighted in yellowish brown, while the lungs are coloured blue and ribs greyish.

In hospitals with a paediatric population it is helpful to make the scan room more child-friendly with special furnishings, stuffed animals or even painted CT-scanners (Figure 3). In our experience, allowing the parents to be present in the CT room during the procedure is much easier for the children. Their presence is dependent on country and local regulations and legislation, and anyone in the room who is not the patient needs to wear a lead apron to protect them from the x-ray radiation. The use of lead shielding to cover parts of the children that are not being imaged is often not useful because it can create artefacts on their images, and can even raise the dose when not used in the proper way.

For most CT exams a contrast agent is needed to create more contrast between organs, vessels and small structures when they only have slight differences in radiation absorption, for instance in the chest and abdomen. An amount of contrast (called a contrast bolus) is injected into the veins, pumped around the body by the heart and often followed by a salty water injection to give the best attenuation of the contrast and to minimise the amount of contrast material given.

Considerations about radiation dose are important in children because of their greater radiosensitivity. In children, the same radiation dose has the potential to do more harm than a similar dose used in adults. For this reason a team of specialists always discuss whether the benefits of performing a CT scan outweigh the possible risk of radiation. To keep the radiation dose to the child as low as possible, different approaches can be considered by the professionals. This is the so-called ALARA principle: As Low As Reasonably Achievable. One of these approaches can be to restrict the area imaged to the absolute minimum necessary to achieve the objective of the CT exam. Close cooperation between the referring specialist, paediatric radiologist (a medical doctor who is trained in interpreting radiological images in children) and paediatric radiographers (the operators of the CT scan) is needed. Current technological improvements in CT scanners have made it possible to lower the radiation dose to such level that the benefits of the CT almost always outweigh the possible risks of radiation. For instance when a child is transferred to an emergency room after a fall or trauma a CT of the head can be performed; if the scan is normal the child can go home. Without CT a child is normally admitted in the hospital for observation. So in some situations CT can prevent hospitalisation and be better for the child and their family, but suggesting a CT is always carefully considered.

In summary, CT scan technology has been continuously improving over the last decade, allowing the imaging of challenging patients like children. The strength of CT technology is in the ability to visualise complex anatomy and diagnose or exclude diseases. It provides information on the size and shape of the organs and can aid the surgeon in deciding how best to treat a specific disease. Given careful consideration, CT scans can be performed in children with success, allowing fast and accurate diagnosis.



of interest.

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A doll representing a child (C) positioned in a vacuum cushion (A and C) for CT of the chest. The lower left image (B) shows an overview made with the CT scanner to plan the scan of the body region



CT scanner painted with a clown to create a more child friendly atmosphere.

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MAGNETIC RESONANCE IMAGING

BY ØYSTEIN OLSEN

RI (or Magnetic Resonance Imaging) is very useful for radiologists in general because it gives very detailed anatomical images of almost any organ or body part. This is particularly useful in children, where the other imaging techniques fall short. For example, CT imaging of the brain does not provide sufficiently detailed images of the brain, and ultrasound of the abdomen does not provide 3D images for surgical planning.

There are two main reasons that MRI is encouraged: first, the different kinds of tissue often appear quite different on MRI, whereas on CT they often give only slightly different shades of grey; second, bone and gas are not an obstacle to MRI, which they are for ultrasound. The particular advantage for children is that MRI does not use x-rays, and we know that children are more vulnerable than adults to long-term adverse effects of x-rays. But there are disadvantages to MRI. For example, MRI requires more expensive equipment and longer scan times, which means that people need to lie still for a relatively long period of time, which can be challenging for some children.

HOW DOES MRI WORK?

The way that MRI works is rather complex. The short version is that hydrogen atoms (which are fortunately very abundant in the body in water and fat) are tiny, weak magnets. Because all of these tiny magnets normally point in random directions, they normally cancel each other out and do not exert any effect. However, when a person enters the tube of an MRI scanner, which is a very strong magnet, all of these little magnets will line up in one particular direction.

The MRI scanner then sends radio waves into the now-magnetised person, and these radio waves make those little hydrogen magnets wobble sideways (through a process called resonance), which in turn make them produce an echo of the radio waves. These these into images.

kidneys working.

DIAGNOSTIC USE OF MRI

Most people think 'brain scan' when hearing MRI mentioned. It is also correct that most MRI scans are of the brain since MRI gives much more information about brain tissue compared to its counterpart, computed tomography (aka CT or CAT scan). Although ultrasound is also a great way of looking at the brain in newborn babies, in older children their hard skulls block the ultrasound waves, making imaging impossible. MRI is very effective for checking whether there is any abnormality in the brain when a child has certain complaints, such as seizures, severe headache or delayed development. Other children may have

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echoes are picked up by antennae or detectors (these are called MRI coils) that are placed close to the body part we want pictures of. The echoes vary depending on 1) from where in the body they are returned and 2) from what kind of tissue they are returned. The scanner can interpret the different echoes and reconstruct

The paediatric radiologist will use the fact that tissues have different MRI echo 'fingerprints', for example to highlight tissues with a high water content - which can indicate inflammation. Sometimes the radiologist will prescribe the use of an intravenous contrast medium (a fluid) which enhances the MRI echo from inflamed or abnormal tissue. As the contrast medium stays in the blood stream for a short while after the injection, this can provide guite detailed pictures of the blood vessels. This can help in diseases where the blood vessels are inflamed (vasculitis) or have developed abnormally, such as in cancerous tumours. Furthermore, because the contrast medium is eventually cleared from the blood by the kidneys, it is also possible to take repeated pictures and literally see the

specific problems, like a brain tumour or epilepsy, for which MRI is crucial when tailoring the best possible treatment for the individual child.

Although problems with joints are not as common in children as they are in adults, arthritis (more specifically inflammation of the lubricating joint lining) is commonly encountered in paediatric healthcare. Left untreated, arthritis often irreversibly destroys the affected joint, and in the worst case, the child may lose the joint function completely. It is easy to understand how devastating this may be for development, education and indeed for the entire adult life. It turns out that MRI is very sensitive to such inflammation, which can often be difficult for the clinical doctor to diagnose at an early stage. MRI can therefore help doctors start the right treatment early and thereby protect children's joints from damage.

MRI of the brain and of joints are only two examples from a wide range of MRI scans offered today. Most of the time an MRI scan is done so that the paediatric radiologist can help his or her clinical colleagues to make the best treatment decision for a child. One important question may be "when should treatment start?" (as in the case of arthritis). Other times it may be "which treatment is the best?" This question is very relevant for a child with cancer. It is so important to give treatment that is powerful enough to kill off the cancer cells. However, since most cancer treatments also can damage normal tissues and organs, it is equally important that it is not too powerful. The radiologist who interprets the MRI scan can help the oncologist balance the treatment, for example, by describing which organs are affected by a tumour and also how a tumour reacts after treatment has started. Surgeons need to plan an operation to be sufficiently extensive to, for instance, remove as much diseased tissue as possible; at the same time they need to spare as much of the normal tissue as possible. MRI is over-


all the best imaging technique to help the surgeon achieve this goal.

MRI – THE EXPERIENCE

For children and their families, the MRI environment is unfamiliar. The scanner itself is shaped rather like a cylinder with a narrow tunnel into which a sliding table brings the person to be scanned. In a typical MRI scanner, this tunnel is about 60 cm (2 ft.) across and 160 cm (5 ft.) long. The MRI coils mentioned earlier are placed around the body parts to be scanned before the table slides in. Sometimes, when pictures of the entire body are needed, coils need to be wrapped around the person top to toe. Many people find this rather uncomfortable. It does not help that the scanner makes guite a lot of noise as it works and that the scan could take more than an hour. Hospitals and departments dedicated to children's needs have made huge efforts, working with scanner manufacturers, to reduce the mental stress an MRI scan may entail. Special mood lighting in the room and inside the scanner and noise reducing technology are examples of this work. The most important job falls to the local team of radiologists, radiographers (technologists) and allied staff in carefully preparing the child and their family for the scan, sometimes using play therapy and a mock scanner. A close family member will usually be allowed to be with the child in the scanning room use of x-rays.

during the entire scan, and most departments have entertainment equipment built right into the scanner, both video and audio, so that the child can watch video or listen to music or perhaps an audiobook during the scan.

These steps may of course not be effective when the patient is an infant or a young child. A common strategy with neonates is to sleep deprive and starve them for some hours, then feed them, swaddle them and hope they will sleep long enough for their scan to be completed. However, the majority of infants and young children require general anaesthesia, which is a considerable challenge in paediatric MRI because it takes up additional time and resources, and a recovery period is necessary after the anaesthetic.

CONCLUSION

MRI is an advanced imaging technique that uses expensive equipment. It requires dedicated contributions from several expert professionals, especially in children because of the long scan times and the not-so-inviting environment of the scanner. MRI therefore needs to be used wisely, which usually means to help answer specific clinical questions. Its major appeal is based on the fact that it can produce very detailed pictures of tissues and organs without the



A young child with two tumours of the right kidney. MRI images help to guide the surgeon so that only the upper part of the kidney (containing a large upper tumour (arrows) and a smaller second tumour in the middle of the kidney (arrowheads)) is removed, avoiding damaging the big renal artery (A) and vein (V) during the operation.

CHAPTER 2: PAEDIATRIC IMAGING TECHNIQUES







MRI image of a young child shows a large tumour (neuroblastoma; arrows) at the back. MRI is used to highlight the tumour sitting around the main artery (aorta; Ao), and between the right kidney (RK) and left kidney (LK) so as to avoid damaging these during the operation. Note that the left kidney (LK) is darker than the right kidney (RK), because its blood supply is disturbed by the tumour mass.



HYBRID IMAGING

BY FRANZ WOLFGANG HIRSCH

paediatric medicine are provided by or significantly influenced by modern imaging. For this purpose, paediatric radiologists have a wide variety of imaging methods available, including ultrasound, x-ray, magnetic resonance imaging (MRI) and computed tomography (CT).

But why is there this range of methods in radiology? Each method is usually used individually, i.e. on its own, and can give very accurate diagnoses. But each method can often only depict certain individual aspects of the underlying illness, rather than the whole picture.

For example, the typical calcified structures of a childhood bone tumour can be displayed much better using normal x-rays than with MRI. On the other hand, MRI is better at displaying the extended soft parts of the tumour and thus over which body parts the tumour extends. This soft tissue is important for the subsequent operation, which must take into account the limits of the bone tumour and also observe safety distances. Both methods thus belong together.

t is estimated that around 70% of all diagnoses in In other cases, such as enlarged lymph nodes or tumours which are more difficult to diagnose, MRI may be used to make better assessments of the tumour type and size. However, knowledge of the function or the metabolic activity may be required, which can be provided using numerous nuclear medicine techniques.

> Until now, it has been necessary to perform these examinations one after the other, in what is called a 'sequential' imaging strategy. Improvements in technology now mean that methods that provide complementary information can now be united in one piece of equipment. This equipment, which provides two or even more methods at the same time, is called 'hybrid equipment'.

HYBRID INFORMATION

Not every conceivable equipment combination makes sense, including from an economic viewpoint. Hybrid equipment is most useful when the information gained presents two completely different, complementary aspects of disease. It would thus only be of limited help to unite two morphological techniques (showing



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Figure 1 shows the fusion principle of PET/MRI: the MRI image (left) and the PET image (right) are recorded at the same time and merged (centre).

form and structure) which can be performed easily one after the other, such as x-ray and MRI. In contrast, over the last ten years, combination equipment has proved to be very useful where it depicts anatomical and functional (or biochemical) information for the illness in one image. The anatomical information is shown in a familiar black and white image, and the biochemical information is superimposed in colour over this grey image. This superimposing process is called *imaging fusion* (Figure 1).

PET/CT

The combination of positron emission tomography (PET), which is a nuclear medicine examination, and computed tomography, is the most frequently used current hybrid imaging technique. Functional and biochemical processes are visualised with the nuclear medicine PET examination and this important information about the metabolic activity of a tumour is superimposed over a highly detailed CT image. During a single PET/CT full-body examination, both the main tumour and any possible metastases can be found. This new hybrid imaging concept has made such an improvement in imaging diagnosis that PET/CT was awarded the 'Medical Innovation of the Year' in 2000.

With various substances that have an effect on metabolism (tracers) and the radioactive markers (radionuclide) linked to these, different tumours can be imaged in a highly specific way and the activity of a tumour may be assessed. Response to chemotherapy treatment can now be visualised by monitoring for a decrease in PET activation in the tumour. It amounted to a medical revolution that you could depict all of this in one single 'hybrid' examination.

However, there is one aspect that limits the innovative PET/CT method problematic for small children, which is radiation exposure. This radiation exposure does not come mainly from the radioactively marked tracer substances in PET/CT, but primarily from the full-body CT examination. Thought must therefore be given to alternatives with less radiation exposure.

PET/MRI

As MRI has significant advantages over CT in children, it would be preferable to use MRI for anatomical imaging together with PET for functional imaging. However, the precise nature of the MRI imaging field meant that this was a significant technological challenge. New PET detectors were required before the first PET/MRI hybrid device was established in 2010.

For children, this development was of great benefit, as children are particularly sensitive to radiation, and they still have a long life ahead of them during which long-term side effects could emerge. The radiation exposure for a paediatric radiological PET/CT examination in 2010 was on average 24.8 mSv (as a comparison: the average annual exposure from natural sources in Europe is just 2.4 mSv). The isolated radiation exposure of the PET components that are still used for PET/MR is just 4.8 mSv, and it is becoming much lower¹. It was thus possible to reduce the radiation dose for PET/MRI by far more than 80% in comparison with a PET/CT full-body examination.

However, at the same time, there are also other, method-specific benefits, which are based on MRI. With MRI, all soft organs can be depicted in particularly high contrast, far better than with computed tomography. This is of great importance, especially for childhood solid tumours. The tumour borders are better determined with MRI. By administering contrast agents intravenously, the blood vessels can also be well imaged. MRI provides images with spatial resolution in millimetres or less, whereas PET provides images with spatial resolution in centimetres, and thus the methods are truly complimentary. This hybrid equipment thus combines the best properties of two worlds: the



Figure 2 shows a boy with a testicular tumour. All metastases (red) are discovered in an examination. In this example, metastases are present in the mesenteric lymph nodes, in the liver and left supraclavicular lymph node (A: MRI image, B: PET/MRI image).

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Figure 3 shows a one-year-old child with a huge tumour in the abdomen and in the spinal canal. The different retention of the PET tracer (red) indicates tumour parts with different activity. Histologically, it was also a tumour made up of malignant neuroblastoma parts and benign ganglioglioma parts.

detailed anatomical world of MRI diagnostics and the functional and biochemical information of nuclear medicine.

PET/MRI IN CHILDREN

The first publication from 2013, which reported on the application of an integrated PET/MRI scanner in children, was able to show that the benefits of both methods can be ideally united, especially for children with lymphoma². In most cases, deoxyglucose is used as a PET tracer, to which a radioactive fluorine molecule is attached (FDG-PET). In childhood lymphoma, the precise overlaying of the PET data produces a significantly improved and more correct stage classification (which in turn influences treatment), even for very small and thus seemingly unremarkable lymph nodes (Figure 1).

In the case of solid childhood tumours, PET/MRI represents an ideal method of identifying the active tumour parts within large tumours (Figures 2 and 3).

This also applies to multifocal brain tumours. Using specific tracers (such as carbon¹¹ methionine), the parts of the tumours with an effect on metabolism, known as hot spots, can be identified. The data is then transferred to the neurosurgeon's 3D navigation system. With a 3D navigation robot, a biopsy can be taken from the location that stands out the most (hot spot biopsy) (Figure 4).

Often following treatment of childhood tumours, a small amount of tissue remains that is mostly only scar tissue from the dead tumour. Unfortunately, the radiologist cannot make the decision as to whether there is still a small remnant of active tumour tissue in this using MRI image alone. In such cases, the superimposed PET image is a great help in making this decision. If there is no longer any sign of radionuclide accumulation, it can be assumed that all the active tumour has gone.

THE OUTLOOK

It is likely that there will be further advances and advantages to PET/MRI in the future³. There are also applications outside the field of paediatric oncology. For instance in epilepsy, when there is no specific brain abnormality seen on MRI, with PET/MRI, one often finds a local metabolic increase, or hypometabolism between seizures.

PET/MRI will become established as a standard method of hybrid imaging in children. The relatively higher costs and the longer examination time of PET/ MRI will be accepted in order to gain the huge benefit of low radiation exposure in children. PET/MRI has thus already become an established paediatric radiological hybrid imaging technique, with highly skilled paediatric radiologists able to manage and interpret the images correctly.

Other developments will concentrate on further reducing the already low radiation exposure of PET components. Future hybrid systems could even do without any radiation: the first successes have been achieved in replacing the biochemical information of PET with new yet just as specific 'MR labelled markers'. This would be a further significant step towards a child-suitable, specific and evidence-based hybrid imaging technique.





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Figure 4 shows the example of a hot spot biopsy in the brain. The hyperintense lesion in the right cerebellum was one of several tumour localisations (A). In the PET, the highest accumulation was found here (B). The data is then transferred to a neurosurgical navigation system and a stereotactic biopsy can be performed in this way at the suitable location (C).

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INTERVENTIONAL RADIOI OGY

BY ALEX M. BARNACLE

nterventional Radiology (IR) is a medical sub-specialty of radiology that uses image-guided keyhole surgery and other minimally-invasive techniques to diagnose and treat a wide variety of conditions in adults and children. It first developed as a medical specialty in the 1960s. Charles Dotter, widely known as the 'father of interventional radiology' pioneered many of these techniques and was awarded the Nobel Prize for his work in 1978.

Medicine has evolved in a whole host of directions over the last 40 years but many would say that IR has been at the forefront of this, introducing the concept of minimally invasive procedures and innovating at an extraordinary rate. The first interventional radiologists developed very fine plastic tubes (called vascular catheters) and metal stents to treat peripheral arterial disease in adults and used real-time x-rays in the IR operating theatre to 'see' these catheters and stents in the bloodstream as they threaded them around the body to the site of disease. It became clear that these basic techniques could be applied in other parts of the body to treat a wide range of conditions. IR quickly went on to develop treatments for conditions such as

gastrointestinal tract bleeds, bile duct disease, liver tumours, bone fractures and kidney stones. Much of IR remains centred on the vascular system, where radiologists can treat aortic aneurysms, abnormalities of the blood vessels of the brain, narrowed kidney arteries causing high blood pressure, and complex vascular malformations or birthmarks. Today, many conditions that once required surgery can be treated non-surgically by interventional radiologists, leading to shortened hospital stays and better outcomes for patients.

It has taken a long time for these techniques to be applied to diseases in children and in many ways paediatric IR is still only beginning to be appreciated. Many of the original techniques developed by radiologists for adult patients had to be adapted to much smaller vessels and organs. Manufacturers have had to match these innovations with significant changes in the size and capability of medical equipment to make it physically possible to treat small children and babies. Traditionally, IR uses x-rays to guide procedures; paediatric IR has had to work with the medical industry to reduce radiation dose during procedures and develop new, safer forms of image-guidance¹



Ultrasound image of a rounded tumour in the neck (*) of a child showing a biopsy needle placed into the centre of the mass.



narrowed airway.

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FIGURE 2



Angiogram of the right kidney in a 12-year-old child. The vascular catheter can be seen in the main artery to the kidney (arrow).

Chest x-ray showing a metal stent (arrow) holding open the trachea of a one-year-old boy born with a

These days, ultrasound is used as much as x-ray to quide what paediatric IR does.

The challenge of IR is to be able to find minimally invasive ways of diagnosing and treating an ever wider range of diseases so that procedures are safer and so that children have a shorter hospital stay and find their treatment easier to cope with. Paediatric interventional radiologists could be asked to treat any number of different conditions in one day, so we need to keep on top of all the latest developments in paediatric medicine and understand innovations in adult IR, so we can take advantage of and modify some of those techniques for children.

Paediatric IR offers a wide range of diagnostic techniques that allow prompt diagnosis for sick children so that treatment can start early. These include sampling or biopsy of organs to diagnose tumours, infections and diseases such as kidney failure or liver disease. Biopsies are often performed under ultrasound guidance as most organs can be well visualised using this imaging modality. In this way, we can avoid exposing the child to radiation. Occasionally, x-rays, CT or MRI are needed to image more deep-seated structures in the body. Imaging allows a radiologist to clearly target the affected organ, acquiring high-yield samples of diseased tissue through tiny coring needles without damaging the nearby vital structures of the body (Figure 1). All of this can be achieved through a very small incision in the skin, usually too small to need stitches or an overnight hospital stay.

Other diagnostic techniques involve the use of contrast medium injected into blood vessels, bile ducts or the gastrointestinal tract to demonstrate abnormalities of those systems. Injecting contrast medium into blood vessels is termed angiography. This technique can be used to obtain a 'roadmap' of the normal circulation supplying a diseased organ such as the blood supply to a kidney tumour. The roadmap can then be studied by surgeons, who use it as a guide to operate safely on that tumour. More innovatively, basic angiographic techniques can be extended to deliver chemotherapy or other drugs directly through the same angiography catheter to the centre of a hardto-reach tumour or to block off the blood vessels supplying a tumour. This may stop or slow the tumour's growth and avoid an operation.

Angiography can also be used to study blood vessels that are inherently abnormal in themselves. Diseases of blood vessels can cause strokes, high blood pressure, dangerous weakening of blood vessel walls and complex birthmarks. By studying the blood vessels with angiography, these conditions can be detected in ways that are often impossible with other types of imaging, partly because the blood vessels are so small (Figure 2). In many cases, the conditions can also be treated, using balloons to stretch up narrow arteries, stents to hold them open or glue to block them off. In trauma scenarios, IR offers emergency life-saving interventions, getting to the source of a bleeding artery very guickly and closing it off from the inside in a quicker and more accurate way than open surgery usually can

IR is central to the management of many other conditions, too. Outside of the cardiovascular system, paediatric IR uses image guidance to treat an extraordinary range of conditions such as joint disease (arthritis), abscesses, kidney stones and blood clots. Real-time imaging in the IR operating suite allows interventional radiologists to place needles into the centre of small, complex joints and into the lining of tendons with immense accuracy, facilitating the injection of steroids to treat arthritis and other inflammatory conditions. In a similar way, deep-seated abscesses can be drained, the urine outflow from obstructed kidneys can be diverted and kidney stones can be lifted out through open the kidney².

from an IR specialist.

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small keyhole tracks into the kidney, avoiding the radical surgery usually needed to remove them by cutting

Paediatric IR also has a key role in simply supporting children's health while they are undergoing other treatments in hospital. IR places gastric feeding tubes for supplemental feeds in unwell children, sites central venous catheters for the administration of intravenous drugs, nutrition and chemotherapy and helps to manage complications of treatment such as blood clots and infection³. In a modern paediatric hospital with an active IR department, it is now very rare for any child to be a patient for very long without receiving support

Specialists from all fields of paediatric medicine increasingly find that they need to work with IR to get the best for their patients. Interventional radiologists are key members of any multidisciplinary

team, advising on imaging findings, diagnostic procedures and novel treatment options. Much of this follows on from advances in adult IR but as paediatric IR matures as a specialty, it is finding new solutions to old problems and exploring a host of new possibilities. Chemotherapy delivered straight into the artery of a retinal tumour has transformed outcomes for children with tumours of the eye (retinoblastoma) who may otherwise have been necessarily rendered blind by conventional therapy. Vascular balloons that can also administer drugs can allow the treatment of blood vessel wall inflammation from the inside. Biodegradable tracheal stents hold babies' narrowed airways open until they have time to grow (Figure 3). IR is a field finely balanced between science and hands-on intervention. As it grows, it offers children the opportunity of faster diagnostics, less painful treatment and quicker recovery times, and it challenges paediatricians to continue to push the boundaries of modern science.

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RADIOTHERAPY IN PAEDIATRIC ONCOLOGY

BY KARIN DIECKMANN AND ANDREA RICCARDO FILIPPI, ON BEHALF OF ESTRO, THE EUROPEAN SOCIETY FOR RADIOTHERAPY & ONCOLOGY

GENERAL INTRODUCTION

Malignant tumours in childhood and adolescence are rare diseases; about 10 out of 100,000 children under the age of 15 develop a malignancy. Due to the combination of treatment options like surgery, chemotherapy, and radiotherapy, long-term overall survival is excellent. Curative therapy for childhood cancer has significantly improved in the last two decades; at present more than 80% of all children affected with cancer are likely to survive for 15 years or longer after diagnosis.

The most common tumours in childhood are leukaemia (Acute Lymphatic Leukaemia/Acute Myeloid Leukaemia), brain tumours (e.g. medulloblastoma, ependymoma, Primary Neuro-Ectodermal Tumours), Sarcoma (Ewing's sarcoma, Rhabdomyosarcoma, Osteosarcoma), Hodgkin's and non-Hodgkin's Lymphomas, Neuroblastoma and Wilms tumours. The distribution of the different types of cancer varies with age, for instance, the peak of incidence of leukaemia is between the ages of one to four, with only a small number of new cases of leukaemia occurring between the ages of 15 and 19.

WHAT ARE THE DIFFERENCES BETWEEN CHILDREN AND ADULTS?

The cancers which affect children and young adults are totally different from those that affect older adults and the elderly. Whilst adults suffer mostly from prostate, breast and lung cancer, children's cancers are leukaemia, lymphomas, brain tumours and sarcomas. But there are other important differences; a life expectancy of 50 years or longer is not unusual for children and adolescents and these patients could therefore experience long-term side effects of treatment. As the physical, mental and sexual development of children is incomplete at the time of treatment, they may suffer from growth or intellectual impairment, hormonal problems and may develop secondary cancers in spite of surviving their childhood cancer.

The goal of treatment in paediatric oncology is therefore not only to cure the disease, but also to reduce life-threatening long-term side effects. This places considerable demands on the paediatric radiologist, surgeon, oncologist and radiation oncologist, who need to work closely together in order to provide the best cancer treatment for children and young adults.



THE ROLE OF RADIOTHERAPY AND TECHNICAL CONSIDER-ATIONS REGARDING WHICH TREATMENT TECHNIQUES ARE AVAILABLE AND ADEQUATE FOR CHILDREN

Treatment of childhood cancers usually involves a combined approach, with the integration of surgery, chemotherapy and radiotherapy. Sometimes radiotherapy can be minimised or even avoided, such as in leukaemia and lymphomas, but it does play an important role in treating the main tumour site in various diseases. When radiotherapy is used to control local disease in solid tumours, high doses are often required. Consequently, the paediatric radio-oncologist makes

a specific technical effort to treat the tumour while simultaneously avoiding affecting surrounding tissue. Magnetic Resonance Imaging (MRI), multi-slice Computed Tomography (CT), and when available, Positron Emission Tomography (PET, used in conjunction with MRI as PET/MRI), are the basis of an accurate anatomical localisation of the cancer

Some special fixation systems may help to optimise the position of the child during treatment planning and may allow a reduction in the size of the target volume. Particular techniques can also help to reduce the volume of tissue irradiated and thus the exposure to organs surrounding the tumour. One example is the use of online image control at the treatment machine, the so-called 'Image-Guided Radiotherapy' or IGRT. A newer technique called Intensity Modulated Radiation Therapy (IMRT) is a sophisticated variant of 3D radiotherapy, which allows seven to ten beams to be applied to the tumour from all directions. The use of these treatments is carefully balanced by radiation oncologists with a special interest and experience in paediatric oncology, as there is a small risk of developing cancer later in life, although this risk is very difficult to estimate or predict accurately.

Newer treatment options are promising, such as proton therapy for children. This unique type of radia-

tion treatment, using particles and not x-ray photons, allows curative doses to be given to the patient while significantly sparing normal tissue due to a physical phenomenon called the Bragg's Peak effect. This effect minimises the irradiation of normal organs that are outside the edges of the tumour. The technique is as good as (or better than) IMRT, but without paying the price of exposing normal tissues and larger regions of the body to low doses. With an increasing number of proton therapy centres worldwide, its use is likely to increase in paediatric oncology. The expectations with proton therapy, especially for specific tumours (such as some brain tumours), are better local control without an increased risk of long-term side effects and secondary tumours. The goal is always to improve the quality of life of children as long-term survivors.

Brachytherapy, a type of radiotherapy using radioactive sources placed inside the tumours or in close proximity through various methods, has relatively few indications in paediatric oncology, and is performed only in specialised centres. The advantage of this technique is to avoid the irradiation of normal tissues by directly treating the tumour (with almost no radiation dose beyond the tumour). One of the most common restrictions to brachytherapy is that tumours are often located in anatomical positions that are difficult to access for radioactive source insertion.

Long-term side effects in children are generally dependent on the dose and volume of radiotherapy received. However, many factors other than the pure dose distribution inside the body should be evaluated by radiation oncologists when estimating the risk of late effects. For example, age itself and the stage of development of the cancer play important roles in evaluating the probability and severity of radiation-related side effects. Like a surgeon, the radiation oncologist is generally able to predict the probability and grade of a specific side effect before the start of radiotherapy; in most cases, according to which structures are included within the planned target volume. The knowledge of the tolerance of normal tissue of different organs, from several radiobiological studies and clinical observations, is essential. At an experimental level, as the data are not yet confirmed, estimations of the incidence of second malignancies may be obtained by mathematical models, taking into account the dose distribution in specific organs and the child's age. However, this approach is not widely accepted and should be further developed before its introduction in clinical practice.

Detailed knowledge of the most sensitive normal structures in children (for example in the brain) and the

Paediatric Imaging

PREDICTION OF SIDE EFFECTS

use of modern techniques in combination with modern imaging are prerequisites to performing an optimal treatment, this should minimise long term side effects. However, because nearly all children get a combined treatment, long-term side effects are not only caused by radiotherapy but also by all treatments combined, and the possible interplay of effects and interactions should be taken into account in the clinical decision-making process. In paediatric oncology it is vital that treated children are followed up and monitored for a long time by various specialists in order to make diagnosis at an earlier stage of a specific treatment-related disease and prevent its possible development.

FUTURE DEVELOPMENTS IN PAEDIATRIC RADIATION ONCOLOGY

Because more than 90% of children are treated according to international guidelines or enrolled in national and international studies, the use of radiotherapy has decreased. There has been a reduction in dose and volume and often a reduction of chemotherapy as well.

As an example, in cases of acute lymphatic leukaemia, prophylactic whole brain irradiation is no longer used,

and only patients with spinal fluid involvement at time of diagnosis or late responders with high-risk features currently receive low-dose prophylactic irradiation. The incidence of behavioural disorders and growth problems can therefore be substantially reduced.

Another example is paediatric Hodgkin's disease, where the number of children treated with radiotherapy has been reduced from 90-100% in the 1990s to 20% in the new ongoing European Paediatric Hodgkin's trial (EuroNet-PHL-C2 Study), where only patients with PET positive lymph nodes receive radiotherapy.

However, radiotherapy is still an important therapeutic tool, and fortunately, it has been adapted. Targets have become smaller, conformal radiotherapy (where the radiation beam is shaped to match the tumour) has become the gold standard worldwide, based on modern imaging such as multi-slice CT, PET/CT and PET/MRI, and the field and doses can be adapted depending on the response of the tumour to chemotherapy or surgery. The continuous progress in terms of better imaging for tumours and organs at risk, better planning using highly conformal techniques or proton therapy, and better prediction of late effects may reduce treatment-related morbidity in the future. To this end it is recommended that the treatment of paediatric patients is restricted to centres with large experience and with radiation oncologists dedicated to radiotherapy for children.

Over recent years, international scientific societies such as the European Society for Radiotherapy and Oncology (ESTRO) or the Paediatric Radiation Oncology Society (PROS) have established platforms for teaching and exchange of experiences in paediatric radio-oncology. Especially in fields of very rare diseases, such as childhood tumours, these platforms are very helpful for the discussion of individual cases and for establishing contact between paediatric radiation oncologists from all over the world.

CONCLUSIONS

Radiation oncology plays a small but important role in paediatric cancer treatment. To get the excellent results achievable in paediatric oncology in terms of survival, with long life expectancy for most patients, highly qualified paediatric radio-oncologists are required. With a deep knowledge of child oncology and the application of the best treatment options/ techniques, excellent local control can be achieved while significantly reducing long-term side effects. The quality of life of these long-term survivors of childhood cancer improves with well designed radiation treatment, where all organs at risk have been spared exposure to unnecessary radiation as much as possible without increasing the risk of local relapses.

FIGURE 1



Response adapted target delineation in paediatric Hodgkin's disease. A: PET-CT at time of diagnosis with enlarged PET positive lymph nodes. B: Early response PET-CT after two cycles of OEPA (Vincristine, Etoposide, Procarbazine, Adriamycine) incomplete tumour regression. C: Target delineation, CTV (Clinical Target Volume) includes the primary tumour extension in cranio-caudal direction, but adapted in axial extension to the post-chemotherapy residual disease.

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NUCLEAR MEDICINE

BY MARINA EASTY

MEDICINE?

Nuclear medicine is a branch of medical imaging that involves using a tiny amount of radioactivity (a radio-isotope) that is attached to a medicine (a pharmaceutical) to form a radiopharmaceutical. This radiopharmaceutical is introduced into the body and reaches the area of interest, and images are taken with a special camera. It can visualise the function of parts of the body or distribution of disease rather than providing a true anatomical picture of the body. The images are usually not as detailed as those seen in an x-ray or other type of scan. In order to improve the pictures that we get, we can obtain a 3D image by rotating the camera around the patient and we can join nuclear medicine images with anatomical images obtained using Computed Tomography (CT) or Magnetic Resonance Imaging (MRI). These pictures will be more detailed and more easily interpreted by the imaging doctors, and the doctors involved in a child's care (Figure 1).

WHAT IS PAEDIATRIC NUCLEAR HOW DOES THE RADIOPHAR-MACEUTICAL GET TO THE RIGHT PLACE?

The radiopharmaceutical can be drunk, swallowed, inhaled or injected. For example, a very small amount of radiopharmaceutical may be mixed with milk and this can be drunk. The time until the stomach empties can then be measured by imaging the milk using a special camera called a Gamma camera. The radiopharmaceutical will emit Gamma rays, (similar to x-rays) that can be picked up by special equipment housed within the gamma camera, and processed into an image by computer.

WHERE DOES THE STUDY TAKE PLACE?

The examination takes place in the nuclear medicine department of the hospital. A specialised children's hospital may have its own nuclear medicine depart-



A whole-body bone scan was performed to assess whether a significant abnormality of the left knee was the cause of a ten-year-old girl's knee pain. The nuclear medicine scan showed very little abnormality.



ment, or the department may be shared with adult patients in a general hospital. If an injection is needed, local anaesthetic cream can be used to stop the injection hurting. For some studies, a child will need to wait for a few hours after receiving an injection, to allow the radiopharmaceutical to spread within the body. In other studies, the child will receive the injection while lying on the camera bed.

When the images are being obtained, the child lies on the Gamma camera. There may be studies where the camera will be placed above the patient over the area of interest or in some cases the camera will rotate around them to obtain a 360 degree view; this will tell the doctor where the area of interest is in the body. The pictures can take from ten minutes to over an hour to obtain. In most cases, we can distract children and babies with feeds, toys, games, music and films in order for them to lie still for the investigation. They are held in place with Velcro straps and padding around their sides, to get the best possible images. Babies usually fall asleep, and older children may need sedation or a general anaesthetic to obtain the best quality pictures. The images may need to be repeated, for example before and after emptying the child's bladder.

Some studies involve looking at the radioisotope travelling through the body, such as a MAG3 renogram, which shows the radioisotope being taken up and draining from the kidneys. This is a dynamic study (Figure 2). Some studies involve static or stationary images only, such as a DMSA (dimercaptosuccinic acid) scan, where the kidney takes up the radiopharmaceutical and shows the outline and function of each kidney (Figure 3).

WHAT KIND OF DISEASES CAN NUCLEAR MEDICINE BE USED TO DIAGNOSE?

Nuclear medicine can be used to assess diseases in many parts of the body, but in children it is particularly used to see if the kidneys function well; in the bones to see if there is any sign of increased bone activity, such as occurs in infection; in the intestine to see whether a child has reflux or slow emptying of the stomach; or occasionally in types of cancer where the cancer cells are visualised by taking up particular injected radiopharmaceuticals. The thyroid gland can also be looked at in babies with low levels of thyroid hormone in their blood to see whether the gland is in a normal position and whether it is functioning normally.

WHAT TYPES OF CHILDHOOD CANCER CAN BE DIAGNOSED WITH NUCLEAR MEDICINE EXAMINATIONS?

An example of a childhood cancer where nuclear medicine is used as part of diagnosis and treatment is neuroblastoma, a cancer of primitive nerve cells. The cancer often starts in the adrenal glands and may spread to the bones. About 100 young children, often under the age of two, are diagnosed with neuroblastoma each year in the UK. The type of nuclear medicine scan used in diagnosis is an MIBG scan (named after the chemical iodine-131-metaiodobenzylguanidine). The radiopharmaceutical contains iodine and

FIGURE 2



A 3D bone scan/CT scan fusion image of both knees of the same child as in figure 1. Although the CT scan shows a bone lesion in the lower left femur, there is no evidence of increased radiopharmaceutical uptake on the bone scan, so this is likely to be benign.

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Posterior image from a static renal scan or DMSA scan showing that the right kidney is functionally smaller than the left kidney.

takes 24 hours to show up clearly in the body. The images are taken 24 hours after the child is injected. In some children, nuclear medicine using MIBG may be used to treat the disease by being attached to a type of radioactive iodine with higher energy radiation called Beta radiation, a form of internal radiotherapy.

HOW OFTEN CAN A NUCLEAR MEDICINE STUDY BE PER-FORMED?

The studies are usually repeated depending on clinical need. A DMSA scan is often performed to look for scarring of the kidneys following a urinary tract infection. In order to obtain the best clinical information, the DMSA study is done six months after the infection has been treated so that the kidneys have had time to completely recover.

HOW MUCH RADIATION IS INVOLVED?

The amount of radiation that a child receives when undergoing a nuclear medicine study is variable, but is normally equal to a few months of normal background radiation.

The amount of radiation that your child will emit after a nuclear medicine study is very low and decreases to almost nothing after 24 hours, however we advise that a partner or friend changes nappies for 24 hours following the study. In addition, you should try not to cuddle your baby until the following day if they have just had a scan. It will be fine to stay in the same room and have meals with them.

KEY MESSAGES

- Nuclear medicine studies are extremely useful to show the area of disease or the function of an organ, for example a kidney.
- 2. The radioactive material is attached to a medicine to form a radiopharmaceutical. The medicine will allow the radioactive material to reach the part of the body where the disease is localised or reach the organ that is being looked at.
- 3. The radiopharmaceutical may be injected, swallowed, eaten or inhaled.
- 4. Gamma rays are emitted by the radioisotope, which can be imaged using a Gamma camera.
- 5. The child may be immobilised on the Gamma camera bed with Velcro straps.
- Each study is different but the pictures can take between ten minutes and several hours to obtain. Some studies need injection first and imaging later in order to allow time for the radiopharmaceutical to reach the area of interest.
- 7. The amount of radiation a child receives is low and is usually similar to a few months natural background radiation.
- 8. The radiation from most nuclear medicine investigations will have gone completely 24 hours after the study.
- 9. Nuclear medicine studies can be repeated as is clinically required or according to a protocol.

"Nuclear medicine can be used to assess diseases in many parts of the body, but in children it is particularly used to see if the kidneys function well; in the bones to see if there is any sign of increased bone activity, such as occurs in infection; in the intestine to see whether a child has reflux or slow emptying of the stomach; or occasionally in types of cancer where the cancer cells are visualised by taking up particular injected radiopharmaceuticals."

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RADIATION PROTECTION

BY GUY FRIJA AND PETER VOCK

INTRODUCTION

The principles of radiation protection in medicine apply wherever ionising radiation is used for diagnosis or therapy. This is particularly important in the case of paediatric patients, because a) children are more sensitive to radiation exposure than adults, as the risks associated with an identical dose decline with age, and b) children have more remaining potential lifespan in which radiation-induced disease, which can have a latency period of decades, may appear.

The principles of radiation protection comprise three elements based on the International Commission on Radiological Protection's (ICRP) system of radiation protection. First, the justification of imaging procedures, which means that an imaging procedure should be carried out only if it provides a benefit greater than the potential harm it might cause. Second, the principle of **dose limitation** specifies that nobody may be exposed to doses that exceed legal dose limits, except in special circumstances such as medical exposure. This principle applies mostly to occupational or envi-

ronmental exposure, which means that patients are exempt from these limits. Lastly, optimisation means that doses should be kept 'As Low As Reasonably Achievable' (the ALARA principle) while maintaining the image quality necessary for accurate diagnosis.

Patients and their parents or carers are understandably concerned about the potential risks associated with radiation exposure. Radiologists and other imaging professionals do their utmost to keep radiation doses at a minimum in order to keep risks as low as possible, while ensuring the best possible image quality for accurate diagnosis. Clearly communicating with patients to foster understanding and alleviate concerns is also part of their duties. It is also worth noting that in general, the risk of radiation is low compared to the risks of inadequate diagnoses or many surgical procedures.

Significant progress has been made in recent years in terms of developing technologically improved equipment, better understanding the risks and benefits associated with radiation exposure through advances in research, and educating healthcare professionals, as

JUSTIFICATION

Justification of imaging procedures is the central task of healthcare practitioners such as clinical radiologists. In Europe, medical exposure to ionising radiation is legally regulated in the EURATOM directives, which aim to eliminate unnecessary exposure, for which the principle of justification is essential (a new, consolidated EURATOM basic safety standards directive will apply from February 2018). The justification of medical exposure to radiation means that an imaging exam must provide a sufficient net benefit compared to the possible detriment the exposure might cause. Only if

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well as in the development of national, regional, and international standards and regulations.

Even though there are higher numbers of imaging examinations being performed every year, it is also true that with modern technology, new research methods, and a greater recognition of the importance of radiation protection among healthcare professionals, patients, policy makers and the wider public, the possibilities for reducing doses while continuing to improve image quality have never been greater.

Making the most of these opportunities and further improving patient safety in paediatric radiology requires multidisciplinary teamwork between radiologists, radiographers and medical physicists, as well as an integrated framework to enhance collaboration in the areas of research, education, infrastructure, advocacy and evidence-based policy making.

Based on this holistic approach, the application of the principles of radiation protection in everyday practice can be further enhanced to ensure patients everywhere receive healthcare of the highest possible guality while ensuring maximum patient safety.

the benefit to the patient or to society outweighs the risks may an imaging procedure be administered.

The process of justification should include the consideration of a variety of factors. These include the appropriateness of the request, optimisation of the imaging strategy, risk-versus-benefit analysis, and consideration of age-specific aspects, for example the avoidance where possible of ionising radiation for children, particularly when frequent follow-up imaging is required, by using alternative non-ionising imaging methods such as magnetic resonance imaging (MRI) or ultrasound.

For effective justification, it is important that referring physicians such as general practitioners, radiologists and radiographers understand their roles in the process and work together as a team. Among the best tools that support doctors in their decision making are evidence-based clinical guidelines that provide referrers with recommendations for most clinical situations. Such guidelines for the use of clinical imaging are produced by a variety of national institutions, medical associations and professional organisations. From February 2018, it will be mandatory for EU member states to ensure that evidence-based imaging guidelines are available to every referrer.

To make imaging referral guidelines easier to use in the complexity of daily practice, the American College of Radiology (ACR) and the European Society of Radiology (ESR) are developing software tools called 'clinical decision support systems' (CDS systems) that allow referring doctors to access the guidelines through a user-friendly computer programme that is integrated into their existing health IT infrastructure. In the United States, where such systems have been in use for a number of years, studies have demonstrated that the appropriateness of imaging exams has improved significantly. The US government has even gone as far as mandating the use of CDS for Medicare and Medicaid

(public health insurance programmes) patients from January 2017. This will also allow for more effective measures to counter self-referrals, which are known to significantly increase the number of imaging exams without clinical justification.

In Europe, the ESR is developing a European system called 'ESR iGuide', based on the American one. This system will provide European doctors with expert recommendations, based on the latest clinical evidence, to support them in providing best-practice healthcare.

OPTIMISATION

Equipment

The concept of optimisation refers to ensuring the quality of the equipment and using it in the optimal way, as well as minimising radiation exposure while maximising image quality. Technological advances and increased competence in using equipment has enabled optimisation in the medical uses of ionising radiation to be continually improved, with computed tomography (CT) being an example where doses have been reduced immensely over the last few years.

The quality and proper use of imaging equipment are essential aspects of successful dose optimisation. In recognition of this fact, the ESR has produced a policy paper on upgrading imaging equipment in Europe. The ESR calls on European healthcare authorities and institutions to establish five-year plans for maintaining and replacing imaging equipment, which is essential in order to keep up with the rapid advances in imaging technology.

Diagnostic reference levels

The concept of diagnostic reference levels (DRLs) was introduced to indicate unusually high radiation doses from medical imaging examinations and procedures. DRLs are a useful tool in the quest to optimise patient doses in diagnostic and interventional radiology. Particular attention should be paid to establishing and using DRLs in paediatric radiology because children are more susceptible than adults to the detrimental effects of radiation. However, despite a large number of studies in European countries, European DRLs for paediatric patients are only available for a few common radiological examinations. The European Commission recognised the need to establish DRLs for radiological examinations and procedures where DRLs are not available, consolidate available information, and provide guidance on what actions are needed in establishing and using DRLs to further enhance radiation protection for children, and approved the 27-month tender project 'PiDRL' in December 2013.

The PiDRL consortium is headed by the ESR. Other participating organisations include the European Federation of Organisations for Medical Physics (EFOMP), the European Society of Paediatric Radiology (ESPR), the European Federation of Radiographer Societies (EFRS) and the Finnish Radiation and Nuclear Safety Authority (STUK) with the Luxembourg Institute of Science and Technology (LIST) as a subcontractor.

The PiDRL project is intended to provide European DRLs for paediatric examinations and to promote their use so as to advance the optimisation of radiation protection for children. The specific objectives are to a) agree on a methodology for establishing and using DRLs for paediatric imaging and b) to update and extend the European DRLs to cover as many procedures as possible.

In the course of the project, a review of literature on patient doses and DRLs for children of different age groups and for different examinations was carried out. Questionnaires were distributed to confirm or update the data on paediatric DRLs in European countries. The review of DRLs has indicated that for interventional fluoroscopy-guided cardiac procedures, no national DRLs exist and that only a few local DRLs have been suggested, while for interventional non-cardiac procedures, no DRLs have been suggested at all.

have not yet been established.

The project's outcomes are consolidated in the document 'European Guidelines on DRLs for Paediatric Imaging'. The draft document was reviewed by stakeholder organisations during a consultation phase in May 2015. Thereafter, the PiDRL Workshop was held in Lisbon on October 15-17, 2015, to present the draft guidelines document to a large audience for comments and review, with almost 200 key stakeholders in paediatric imaging participating in the workshop (radiologists, radiographers, medical physicists, policy makers, etc.). Currently, the PiDRL consortium is finalising the 'European Guidelines on DRLs for Paediatric Imaging'. The final docu-

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The PiDRL consortium strongly recommends that DRLs should be based on patient dose surveys and should sufficiently cover all types of common highdose paediatric radiology practices. The implementation and the results of patient dose surveys, and the subsequent procedures to establish DRLs, should be documented in a way that enables a reliable comparison of DRLs. This will allow trends in their development to be followed-up and possibly established as European-wide preliminary levels where national DRLs ment will be submitted to the European Commission for review and approval at the beginning of 2016.

To find out more, visit www.PiDRL.eu

CONCLUSION

Even though much has been achieved in terms of technological advances and improved competence in radiation protection among medical professionals in recent years, continuing to work for behavioural change to ensure that radiation protection principles are followed by all practitioners in daily practice is important. Through increasing awareness, providing incentives, developing more effective and efficient regulatory measures and by fostering a multidisciplinary approach to radiation protection that also involves patients, parents and carers, radiation protection must continue to lie at the heart of the overall drive to improve quality of care and patient safety in medical imaging. The ESR is pursuing this aim through its EuroSafe Imaging campaign, which was launched in 2014.

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IMAGING THE BRAIN AND SPINE

EPILEPSY AND STROKE IMAGING CONGENITAL BRAIN ANOMALIES SPINE IMAGING



EPILEPSY AND STROKE IMAGING

BY ANDREA ROSSI

INTRODUCTION

The first clinical presentation of a child with epilepsy or stroke is typically a dramatic and frightening event that prompts urgent medical evaluation. It is estimated that 25,000 to 40,000 children every year in the US experience their first seizure. Although stroke is less frequent, affecting approximately 3,000 children and young adults per year in the US, its consequences may be devastating, with a 20% to 40% mortality rate and a 60% chance of permanent neurological problems in survivors.

There is a common misconception that seizures and epilepsy are the same disease. In fact, having a single seizure is not necessarily considered to be epilepsy, and children with epilepsy will typically experience multiple seizures over a long period of time. On the other hand, recognition of stroke is often delayed or even missed in children, and many children with stroke symptoms can be misdiagnosed with more common mimicking conditions, including epilepsy. Thus, getting the right diagnosis at the beginning is of crucial importance for correct management, so that the right treatment and ageappropriate rehabilitation are put in place to minimise long-term functional impairment or disability in survivors.

Among the various diagnostic tests that are available in the emergency department, brain imaging plays a pivotal role in the early diagnosis of patients with epilepsy or stroke. There are multiple factors that influence the timing of imaging and the choice of the most appropriate method, including the severity of the presentation, patient age and radiation exposure concerns. Magnetic resonance imaging (MRI) has a key role both in emergency situations and as a routine imaging method; however, computed tomography (CT) is still useful in emergency, especially in unstable patients. Ultrasound provides an excellent bedside alternative for the non-invasive evaluation of newborns and infants.

IMAGING EPILEPSY

The presentation of a first seizure in a child is often a dramatic event that prompts urgent medical evaluation. In this context, neuroimaging studies are often requested. New-onset seizures presenting with evidence for a medical emergency, such as increased

IGURE 1





Type IIb focal cortical dysplasia (FCD) in a four-year-old girl with a left frontal EEG abnormality. (A) Axial T2-weighted MRI image and (B) axial FLAIR image show subcortical abnormalities (arrow). (C) Reformatting a 3D MRI sequence shows an area of grey-white matter blurring (circle). More detailed sequences (D-F) all show the cortical dysplasia (arrows) which was confirmed at surgery

CHAPTER 3: IMAGING THE BRAIN AND SPINE





intracranial pressure or status epilepticus, always need emergency imaging. In some of these cases, computed tomography (CT) is performed, despite its use of ionising radiation and its well-known relative insensitivity to brain abnormalities compared to MRI, because of the wide availability and scanning speed that may avoid the need for sedation. However, once surgical emergencies have been ruled out, MRI is often performed to identify possible structural causes. It is important to realise that MRI is not always necessary; for instance, certain kinds of epilepsy in children and adolescents do not require imaging studies.

Drug-resistant epilepsy, where the seizures do not respond to a single or combination of anti-epileptic medication, is a chronic debilitating disorder, but many cases are potentially curable by surgery. Neuroimaging plays a crucial role in the workup of patients with drug-resistant epilepsy, as it may reveal a potentially treatable underlying cause. However, many small anatomical abnormalities, most notably focal cortical dysplasias (FCDs), may be extremely difficult to identify on conventional MRI studies. The principal role of neuroimaging studies is to identify both the epileptic zone that must be resected, as well as the neighbouring brain areas that must be preserved to avoid neurological defects after surgery. Imaging also helps to identify children with other medical conditions that require intervention or evaluation of other organ systems, such as tuberous sclerosis, for instance.

It is important for paediatric radiologists to recognise that patients need slightly different specific MRI scanning techniques according to their age, particularly as brain development is not yet complete in very young children. Routine administration of gadolinium contrast material during scanning does not help in children with epilepsy, and should be reserved for cases where other diagnoses are suspected.

IMAGING STROKE

Stroke is defined as the rapid loss of brain function due to a decreased cerebral blood flow, and can be classified into ischaemic and haemorrhagic. Arterial ischaemic stroke (AIS) is more frequent than haemorrhagic stroke (HS) in children as well as in adults, but the proportion of HS is higher in children than adults (45% versus 20%). Because stroke is relatively uncommon in children and making an accurate diagnosis remains difficult, often taking longer than it does to recognise the problem in adults, many cases remain misdiagnosed. Acute ischaemic stroke in neonates may have very few symptoms, mainly presenting with short-lasting seizures that can easily remain undetected.

In older children, strokes typically present more as they do in adults, with face or body weakness. Unlike adults, who may have atherosclerotic blood vessel and heart disease, as well as high blood pressure, children with strokes often have tumours, inflamed blood vessels or inherited conditions as the underlying cause.

When a child is suspected to have suffered from a stroke, CT is typically performed as an emergency. CT can rapidly identify bleeding in and around the brain, as well as the size of the stroke itself. However, MRI has the advantage of several different techniques to evaluate stroke patients, and is usually used for patients who do not need emergency surgery. A specialised type of MRI called diffusion-weighted imaging (DWI) is useful to demonstrate the stroke and size of the neighbouring affected brain areas, before it can be seen by normal MRI techniques. Susceptibility-weighted imaging (SWI) is particularly good for showing fresh and older blood, as well as better characterising the size of the neighbouring brain areas that can potentially recover





arteriopathy of childhood.





Cerebral stroke in a six-year-old boy. (A) Axial MRI diffusion-weighted image and (B) corresponding ADC map show large area of swollen brain in the right middle cerebral artery territory; which is difficult to identify on conventional axial T1-weighted (C) and axial T2-weighted images (D). (E) Axial SWI sequence shows prominent medullary veins (arrowheads) around the stroke area. Detailed imaging of the blood vessels (F and G) show severe reduction of the blood vessels in this area, consistent with a diagnosis of focal cerebral

Other MRI techniques called arterial spin labelling (ASL) have been developed to investigate blood flow in the brain without using a contrast agent (which can need a rapid injection of a large volume of contrast in newborns and young children). These can be used to provide detailed images of the blood vessels to the brain.

CONCLUSION

Paediatric epilepsy and stroke are significant causes of childhood disability. Many challenges exist in the diagnosis and treatment of these children, including the difficulty of recognising the causal factors, delays in the diagnosis, the frequent need for anaesthesia support for diagnostic MRI, and the uncertainty about treatment, especially in stroke patients. Neuroimaging studies are crucial both in the initial workup and in the follow-up; however, the choice of the most appropriate method and the implementation of study protocols must be carefully tailored to address the individual child. The need for expert paediatric neuroradiologists and for a programme of continuous training of those involved in paediatric diagnostic and interventional neuroimaging procedures represents a challenge that the medical academic and scientific community must address if patient care in these important fields is to be improved.





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Cerebral stroke in a 14-year-old girl with moyamoya disease. (A) Axial diffusion-weighted image and (B) corresponding ADC map show restricted diffusion consistent with acute stroke in the left parietal lobe; (C) axial T2-weighted and (D) axial FLAIR images show older abnormalities on the right side. (E) Dynamic susceptibility contrast perfusion-weighted imaging shows an area of reduced cerebral blood flow (arrowheads) corresponding to the diffusion-restricted zone; detailed imaging of the blood vessels (F and G) shows extensive additional abnormal vessels (arrowheads), secondary to severe stenosis of the carotid arteries (arrows).

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CONGENITAL BRAIN ANOMALIES

BY ELIDA VAZQUEZ, IGNACIO DELGADO AND ANGEL SANCHEZ-MONTAÑEZ

INTRODUCTION

Congenital brain anomalies are abnormalities in brain development that are present at birth, although some may not be immediately obvious. They are relatively uncommon, occurring in less than 1% of all births. The severity of these conditions varies considerably from mild forms to serious malformations that lead to significant disability, such as cognitive impairment, speech impediments, and movement disorders.

A variety of genetic and environmental factors are potential causes of the interruption in proper brain development, including inherited or spontaneously occurring genetic defects, maternal infection, use of certain drugs, exposure to toxic substances, or trauma to the unborn foetus. Because most brain structures develop at about the same time during foetal life, it is common to see several associated anomalies, which we refer to as clinical syndromes.

Congenital brain malformations are usually classified according to embryological timing, as follows:

- Disorders of neural tube development, occurring in weeks 3 to 4 of foetal development and including neural tube defects, myelomeningocoele, Chiari malformations, and cephaloceles
- Disorders in formation of brain ventricles, during weeks 5 to 8, comprising holoprosencephaly and posterior fossa cysts
- Disorders of brain connectivity, in weeks 7 to 20, and covering corpus callosum agenesis, septooptic dysplasia, and septum pellucidum agenesis
- Cerebral disorders, occurring in weeks 12 to 23, which include malformations of cortical development

IMAGING MODALITIES

Dysmorphic features or abnormal neurological behaviour in a neonate or infant may suggest cerebral malformations. Various imaging techniques can be used to evaluate the central nervous system and clarify the severity of the condition. The paediatric radiologist will choose the most appropriate technique to provide the correct diagnosis. Ultrasound is often the initial method used to exclude a major structural anomaly in neonates or infants. The advantages of cranial ultrasound are that it can be performed at the bedside with minimal disturbance to the child, and that sedation is not required. Magnetic resonance imaging (MRI) is preferred to computed tomography (CT) in children because it does not use radiation. It is extremely useful for establishing the diagnosis and estimating the consequent impairment. Moreover, specific patterns seen on MR images can indicate the need to perform genetic studies that will clarify the condition, which is particularly important for genetic counselling in future pregnancies. MRI provides valuable information on the brain architecture and can clearly depict structural abnormalities in a non-invasive manner. In addition, certain advanced MR techniques such as diffusion tensor imaging (DTI) and fibre tractography (FT) can be used to evaluate brain connections, which can suggest how well the brain is functioning. Post-processing programmes are used with the images obtained to visually reconstruct the white matter fibres, and represent them by colour-coded



maps showing the predominant direction of diffusion in different colours (Figure 1).

Nowadays, many brain malformations are diagnosed antenatally. Ultrasound is the modality used in prenatal screening, but foetal MRI should be performed when an antenatal ultrasound examination shows abnormalities, if there is a family history of central nervous system (CNS) malformation, in genetic syndromes with CNS involvement or brain damage, and in cases of in brain injury or a risk of brain damage (maternal infection). Several reports have shown that, for a variety of indications, foetal MR imaging can detect abnormalities that are not seen in up to 50% of ultrasound examinations. MRI of the developing foetal brain allows the radiologist to assess the multi-layered appearance of the brain tissue, the timing of development, myelination status, and changes in ventricular size. Imaging protocols have to be adapted according to the suspected pathology as well as the gestational age. Accurate interpretation of foetal MRI provides valuable information that can help prenatal counselling, facilitate management decisions, guide therapy, and support research studies.

To illustrate the value of MRI examination in paediatric patients with congenital brain anomalies, the following are a few examples of the more common conditions encountered in this population.

CORPUS CALLOSUM ABNORMALITIES

The corpus callosum is a bundle of nerve tissue that allows communication between the two brain hemispheres. Children with callosal abnormalities may have a wide range of symptoms from mild to severe. depending on the type of anomaly (agenesis, hypogenesis, dysgenesis, hypoplasia) and other associated brain abnormalities. Patients with the most severe forms may have intellectual retardation, seizures, hydrocephalus, or spasticity. Callosal defects are usually detected by prenatal ultrasound, but MRI is very important in ruling out associated abnormalities (Figure 1). Several potentially severe syndromes (e.g., Arcadi or foetal alcohol syndrome) involve callosal developmental defects. Hence, this information is highly relevant for pregnancy management.

MYELOMENINGOCOELE AND CHIARI MALFORMATION

Myelomeningocoele and myelocoele are forms of spina bifida, a condition in which the bony spinal canal does not properly close during development, and the spinal cord and meninges protrude through a saclike opening in the back. Myelomeningocoele is often associated with Chiari malformation type II, in which the cerebellar and brain stem tissue herniates to below the base of the skull. After birth, these patients frequently develop hydrocephalus and have orthopaedic and sphincter problems. Although this condition is first identified with ultrasound, based on findings of ing the protein filamin A (Figure 2).

a small posterior fossa cerebellar herniation ('banana sign') or frontal concavity ('lemon sign'), MRI can better demonstrate the amount of herniation and other additional CNS anomalies, such as callosal dysgenesis. Closure of the defect by foetal surgery prevents secondary damage to neural tissue from exposure to amniotic fluid, may reverse the hindbrain herniation, and may improve outcome.

CORTICAL DEVELOPMENTAL MALFORMATIONS

These include a wide range of disorders in migration and organisation of the functioning cerebral cortex and are common causes of neurodevelopmental delay and epilepsy. During normal brain development, neurons (brain cells) form in the periventricular region and then migrate outward to form the cerebral cortex in six onion-like layers. Many genes linked to several pathways are known to regulate neuronal migration, but the mechanisms are still poorly understood. Foetal MRI has significantly contributed to prenatal identification of malformations of cortical development and grey matter heterotopia, particularly during the later stages of pregnancy, when they are usually not seen on ultrasound. The optimum time to perform the examination is after 30 to 32 weeks' gestation. If an earlier examination is performed, it should be repeated after this gestational age.

In **periventricular heterotopia**, some neurons fail to migrate to their proper position at the cortical plate, and instead form nodules along the ventricular walls that are similar in signal intensity to the grey matter on MRI. The condition usually manifests with seizures, often at the time when the patient is a teenager or young adult. Most cases are caused by mutations in FLNA, a gene that provides instructions for produc-





- B:
- C:
- D:



A-D: Partial corpus callosum agenesis

A: Foetal MRI image of a foetus in the 23rd week of gestation referred to investigate the absence of the corpus callosum.

A coronal view of the brain on foetal MRI confirms that there are crossing corpus callosum fibres. After birth, a diffusion tensor imaging (DTI) colour map confirms the partial absence of white matter tracts, with crossing red fibres corresponding to corpus callosum remnants (arrows). After birth, fibre tractography (FT) confirms the partial absence of white matter tracts, with crossing red fibres corresponding to corpus callosum remnants (arrows).





Periventricular heterotopias. Differential diagnosis with tuberous sclerosis complex. Sagittal MRI image from a foetus in the 30th week of gestation shows irregular ventricular margins with small nodules around the ventricles corresponding to heterotopias (A). Subependymal nodules in a foetus with tuberous sclerosis complex are seen on Haste axial image (B).



Polymicrogyria: Axial foetal MRI image of a foetus in the 31st week of gestation, at the level of the lateral ventricles, shows multiple bilateral cortical infoldings, consistent with polymicrogyria (arrows) (A), compared to a normal foetus at the same stage of development (B).

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Agyria/pachygyria results from a disturbance of the migration of the immature brain cells which leads to absent (agyria or lissencephaly) or decreased sulcation or gyri (pachygyria), the normal convolutions or folds of the brain. Patients with type I (classic) lissencephaly typically show markedly decreased muscle strength and limited movement, whereas type II is associated with muscular dystrophy-like syndromes. In type I lissencephaly the brain has a typical hourglass or figure 8 appearance, with a few poorly formed gyri and a smooth outer surface. About 60% of patients with lissencephaly carry genomic alterations or LIS1 gene mutations.

Schizencephaly is a rare disorder that appears as unilateral or bilateral clefts extending across half the brain, from the ventricular surface to the periphery. Patients may show developmental delays, delays in learning speech and language skills, or movement problems. Bilateral clefts elicit more severe symptoms. Although the exact pathogenesis of this condition is uncertain and familial cases have been reported, some authors propose early problems with the blood supply to the brain in utero as the origin. Schizencephaly occurs more often in abandoned or adopted children, possibly because of maternal abuse of cocaine or other drugs that affect the foetus. MRI is the diagnostic imaging modality of choice, enabling identification of the cleft with the abnormal grey matter lining.

Polymicrogyria appears as multiple abnormal infoldings of the developing cortex (Figure 3). Although this condition is often sporadic or genetic, it is also seen secondary to intrauterine viral infection, blood supply problems in twins, and syndromes such as Zellweger syndrome or Fukuyama muscular dystrophy.

CONCI USION

Technical advances in foetal and postnatal MR imaging have made it an invaluable tool for the clinical evaluation of suspected CNS abnormalities. The significant recent improvements have provided uniquely detailed quantitative information about white-matter microstructural organisation and connectivity, with promising applications.

The diagnostic and treatment goals for the affected population can be best achieved through collaboration between paediatric radiologists, obstetricians, child neurologists, paediatric neurosurgeons, and perinatologists.

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SPINE IMAGING

BY WINNIE C.W. CHU

WHEN DO CHILDREN NEED IMAGING OF THE SPINE?

Some children have congenital malformations of the spinal canal and its content. Some of these anomalies are obvious with the presence of a large mass over the lower spinal region at birth. However, some anomalies are hidden (we call these occult spinal dysraphism, OSD). There are usually clues indicating the presence of such spinal malformation, i.e. birthmarks that can be seen on overlying skin. These cutaneous stigmata include sacral dimples, hairy patches and even small soft tissue masses. Imaging is indicated when a newborn is found to have such cutaneous stigmata, to rule out the presence of associated spinal malformation.

In older children, the indications for spine imaging are similar to those in adults. Some children present with acute and severe symptoms such as weakness or paralysis of the limbs, loss of sensation, or loss of bladder and bowel function. Urgent imaging of the spine is important to rule out any surgically correctable causes and to try to prevent development of irreversible damage to the spinal cord. Other less acute indications include back pain, unsteady gait and rotational deformity of the spine (atypical scoliosis). The main aim of imaging is to look for the cause of the symptoms, such as infectious disorders, tumours, trauma, vascular anomalies and inflammatory processes.

WHAT IMAGING MODALITY TO CHOOSE?

Radiographs, though widely available, are insufficient for evaluating the spinal cord and other contents within the spinal canal. However, x-rays are often the first imaging modality used to evaluate the spinal column to look for any bone abnormalities, in particular, to detect bone fractures in children after trauma. Fractures seen on radiography are usually better visualised with CT and MRI, which in addition is useful for evaluating concomitant underlying spinal cord injury.

Ultrasound (US) is a well-established and readily available non-invasive technique for evaluating the spinal cord. It is particularly useful in newborns and infants as the spinal canal can be clearly visualised by US because the bony structures at the back of the spine are not fully formed at this age. Furthermore, US involves no radiation and the infants usually do not require sedation during the examination. For infants presenting with cutaneous stigmata, which may suggest congenital spinal anomalies, ultrasound is the first study to offer. US provides adequate screening with high sensitivity and specificity for low-risk subjects presenting with congenital midline skin lesions other than simple sacral dimples. It is also useful for diagnosing lumbosacral soft tissue masses that have specific US features.

When the infant ages, with progressive ossification of the spinal elements, the utility of spinal US becomes more limited. MRI is the modality of choice for evaluation of spinal canal contents in older children. Although US is the preferred first-line imaging modality for most infants with suspected OSD, MRI is the first-line imaging investigation for infants at very high-risk of OSD, such as those with associated malformation of the bowel and urinary bladder. It is also useful as a second-line imaging method for better anatomical delineation of abnormalities detected by US, which require surgical planning, as well as to screen for associated anomalies higher up in the spinal cord.

MRI is actually the imaging modality of choice for detailed analysis of the spinal canal contents as it provides superior soft tissue contrast resolution relative to other imaging techniques. Similar to US, MRI does not involve ionising radiation. However, MRI is not readily available in clinics or smaller hospitals. Sedation is always required for young children as they need

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to stay completely motionless inside the MRI scanner during the examination to allow for acquisition of quality images.

WHAT WILL THE IMAGING EXAMINATION BE LIKE?

For ultrasound (US) scanning of the spine, infants are preferably scanned facing down, with their bodies curved over a pillow and head slightly elevated. This allows the fluid within the spinal canal to accumulate low down in the canal, hence optimising visualisation. It also enables the lower back to curve more for better defining of the junction between the lumbar spine and sacrum. Alternatively, in order to calm a restless baby, scanning can be done with the infant lying on one side to allow for bottle or breast feeding. Sedation is usually not required during US examinations.

For MRI scanning of the spine, children are preferably scanned facing up, lying on the scanning table. During the image acquisition, the child is required to be motionless and the total examination time is about 30 minutes, therefore sedation is usually required for infants and children under six years old or in those children who cannot obey instructions and stay calm.

WHAT CAN IMAGING OF THE SPINE TELL YOUR CHILD'S DOCTOR?

In infants who have cutaneous stigmata, there may be associated occult congenital spinal malformations, such as tethered/low lying cord, presence of abnormal fat components within the spinal canal (intraspinal lipoma) or abnormal fluid distension within the central canal of the spinal cord (syringomyelia). The above can only be detected by imaging. For children with obvious soft tissue masses in the lower back, there may be abnormal communication between the spinal canal with these masses, for instance, sometimes the spinal cord is exposed to air without intact skin (myelocele myelomeningocele) while some masses are associated with intact skin (lipomyelocele, lipomyelomeningocele or meningocele). MRI is useful to differentiate the different kinds of congenital lesion, to delineate the anatomy for surgical planning, as well as to evaluate other associated abnormalities in the brain and the spine. It is important to detect these congenital spinal malformations by imaging at a very early stage of life, so that early surgical correction can be performed to prevent permanent neurological damage or infection of the spinal cord at a later stage. In general, the surgical goals include removal of any fatty tissue within the spinal canal, identification of any defect in the overlying tissues, release of any spinal cord tethering and preservation of normal nerve elements within the spinal cord.

For children with trauma, if spinal fracture is detected or whenever the victim presents with neurological problems, it is important to perform MRI of the spine to look for any blood clot or broken spinal elements within the spinal canal causing compression of the spinal cord. Prompt surgical removal of the blood clot or broken fragments can help to prevent irreversible damage to the spinal cord and permanent neurological damage to the child. It is important to recognise that traumatic injuries in children differ from those in adults as children have proportionally bigger heads, weak neck muscles, incomplete bony formation and more laxity of the spine. Children are prone to upper neck injuries without bony fractures. We call this kind of injury SCIWORA (spinal cord injury without radiographic abnormality), which can only be detected by MRI but not on radiographs or CT. The neurological consequence can be serious in this kind of injury.

In non-traumatic cases when children present with sudden alarming neurological symptoms such as weakness or paralysis of the limbs, sudden loss of sensation, or loss of bladder or bowel function, urgent imaging of the spine is important to identify the causes of spinal cord abnormality, whether it is surgical (where a mass is present) or non-surgical. In surgical cases, when the cord is compressed by an external





ration on real-time imaging.

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Positioning of infant during ultrasound scanning of the spine. The baby is scanned facing down with his body curved over a pillow and head slightly elevated. This position allows the spinal fluid to accumulate low down within the spinal canal hence optimising visualisation of the spinal cord and other contents within the spinal canal.



Normal appearance of the spinal cord on ultrasound. The lower end of the spinal cord (arrow) is around lumbar vertebral level L1-L2. The nerve roots (arrowheads) can be seen moving with respi-





Ultrasound image showing abnormalities of the spinal cord which is low-lying (arrow) and terminates at a low lumbar level L5. There is abnormal accumulation of fluid (arrowheads) within the lumen of the spinal cord, which appears as a dark line within the cord.





Normal MRI appearance of the spinal cord (arrow), which terminates at the level of L1 and the nerve roots (arrowheads).

MRI showing congenital malformation of the spine with abnormal subcutaneous fatty mass (black arrow) over the lower spinal region, which creeps into the spinal canal through a wide bony defect of the lumbar spine. The cord (arrowhead) is tethered to this abnormal mass. This is a lipomyelocele.

mass, such as a tumour, a collection of pus (abscess), vascular malformation, etc., an urgent operation is necessary to relieve the spinal cord from continued compression. The surgical approach depends on the nature of the compression. If the compressing mass is benign or well defined, complete excision of the mass is usually attempted during the first operation. If the mass cannot be immediately removed completely, usually a bony defect is made on the posterior element of the bony spine (laminectomy) in order to create more space for the spinal canal so as to release compression onto the cord. At the same time, the mass will be excised as much as possible (debulking). Tissue obtained will be examined under microscope to find out the exact nature of the lesion. Some tumours are sensitive to chemotherapy and radiotherapy and follow up MRI can help to monitor

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the sizes of these tumours before definitive surgery is performed.

In non-surgical cases, there may be infection, inflammation (myelitis) or demyelination (such as multiple sclerosis) of the cord, leading to the neurological symptoms. These children do not require surgery. Treatment depends on the cause of the myelitis, but typically antiviral/antibacterial medications or steroids will be given, hopefully to reverse the cause and bring the cord back to normal function. Sometimes, if the treatment is unsuccessful, more aggressive medication such as interferons, immunoglobulins or some chemotherapy drugs will be used. The outcome of myelitis is highly variable depending on the cause and treatment response. Again, follow-up MRI can help monitor progress and treatment response of the disorder.

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IMAGING THE LUNGS AND HEART

PAEDIATRIC LARGE AIRWAY DISORDERS



IMAGING OF $N \vdash R \leq I \mid I \mid$ UNG DISEASES

BY MATTHEW A, ZAPALA AND EDWARD Y, LEE

INTRODUCTION

Childhood interstitial lung diseases (chILD), also often called paediatric diffuse lung disorders, are a diverse group of lung abnormalities that affect the lung tissue surrounding the airways. These diseases present in infants or older children, causing breathing difficulties, low oxygen blood levels and lung abnormalities on imaging studies. Radiology plays an important role in early and accurate diagnosis of chILD in paediatric patients. Here we review the imaging techniques and findings of selected chILD that are encountered in the paediatric population.

IMAGING EVALUATION

Radiology is critical in identifying and managing patients with chILD. Unlike adults, the majority of interstitial lung diseases in children have an underlying cause. Thus, timely and accurate diagnosis, along with appropriate early intervention, is crucial in the treatment of children with chILD. However, as chILD includes relatively rare diseases with nonspecific symptoms, they remain a challenge to diagnose. A multi-disciplinary approach to diagnosing chILD, consisting of radiology, clinical evaluation and pathological correlation together, is superior to any one component alone.

IMAGING TECHNIQUES

In terms of a radiological evaluation, chest radiographs and computed tomography (CT) are currently the most widely used imaging techniques in evaluating chILD. Generally, chest radiographs are the initial imaging test of choice as they are widely image quality.



available, simple to perform and involve a relatively low radiation dose. Unfortunately, the initial screening chest radiographs in paediatric patients with chILD are often normal. Therefore, high-resolution CT (HRCT), which provides increased image detail, may be necessary for the improved detection of subtle and earlier cases of chILD. However, HRCT uses higher levels of ionising radiation than chest radiographs. In addition, in infants and young paediatric patients (five years old or under), sometimes sedation or anaesthesia is necessary to keep the patient still during CT imaging. Therefore, HRCT is only performed on paediatric patients where there is a high clinical suspicion of chILD. Furthermore, the imaging protocol should be adjusted according to the individual patient size, with every effort to minimise radiation exposure to 'as low as reasonably achievable' (ALARA) without sacrificing diagnostic

SPECTRUM OF IMAGING FINDINGS

The imaging findings associated with chILD can be separated into five distinct groups: diffuse developmental disorders, alveolar growth abnormalities, surfactant dysfunction disorders, specific conditions of unknown or poorly understood etiology, and other childhood diffuse lung disorders.

Diffuse developmental disorders

The diffuse developmental disorders category involves diseases that are believed to originate in the womb during early foetal lung development. Patients are usually non-premature babies who present with progressive difficulty breathing and cyanosis (bluish discoloration of the skin due to low blood oxygen) within 48 hours after birth. As these disorders also usually

involve vascular changes in the lungs, affected paediatric patients often develop severe pulmonary hypertension (high blood pressure that affects the arteries of the lungs). Chest radiographs typically show diffuse hazy opacities with decreased lung volumes. Chest radiographs may also show increased pulmonary blood flow and enlargement of the main pulmonary artery if there is concurrent pulmonary hypertension. Affected infants are usually on ventilators and are more prone to problems associated with ventilators including an increased rate of pneumothorax (a collection of air in the chest outside the lung) and pneumomediastinum (air surrounding the heart and central blood vessels).

Alveolar growth abnormalities

Alveolar growth abnormalities are the most common cause of chILD in infants. At the outset of foetal development, these patients have normal developing lungs. However, an external condition or event causes chILD to develop either before or after birth. Prenatal conditions include lung underdevelopment due to too little amniotic fluid or neuromuscular disease. Postnatal conditions include prematurity-related chronic lung disease and structural changes to lung tissue related to chromosomal abnormalities or congenital heart disease. Affected infants usually present in the newborn period with difficulty breathing proportionate to the extent of their disease. The mortality rate of infants with alveolar growth abnormalities is high at 34%. While imaging findings can vary in this category, specific findings related to chronic lung disease of prematurity include reticular (spider web like) opacities, cystic lucencies (clear cystic spaces in the lungs) and disorganised areas of collapsed and aerated lung tissue.

Surfactant dysfunction disorders

Surfactant dysfunction disorders are caused by genetic disorders that either directly affect surfactant itself or

impact surfactant function. Surfactant is a complex of lipids and protein which is produced by alveolar (air sac) lung cells and reduces surface tension in the lung preventing lung collapse and increasing the ability of the lung to expand. Affected patients usually present shortly after birth with findings ranging from mild asthma-like symptoms to respiratory failure. Imaging findings on chest radiographs include diffuse or patchy, hazy, granular opacities similar to surfactant deficiency syndrome of prematurity. CT imaging demonstrates a linear pattern superimposed on a background of ground-glass opacity (shading) which resembles irregularly shaped paving stones known in radiology as 'crazy paving'. If affected patients survive out of infancy they often develop chest wall abnormalities such as pectus excavatum (caved in or sunken chest) likely due to the reduced ability of the lung to expand, due to the lack of surfactant. Surfactant dysfunction disorders are usually diagnosed by genetic testing.

Specific conditions of unknown or poorly understood etiology

Specific conditions of unknown or poorly understood etiology include two entities that are unique to infants: neuroendocrine cell hyperplasia of infancy (NEHI) and pulmonary interstitial glycogenosis (PIG). Infants affected with NEHI are usually term newborns who are normal at birth and present within three months, sometimes following a viral respiratory infection. Their symptoms usually involve rapid breathing, chest retractions, low oxygen in the blood, and crackles heard on physical examination. However, they do not have a cough or wheeze. On chest radiographs, the imaging findings are characterised by increased lung volumes with a variable degree of increased opacities around the heart and great vessels. Characteristic CT findings are geographic ground-glass opacities (shading) seen surrounding the heart and great vessels. CT

FIGURE



Figure 1A: Frontal chest radiograph of a fourmonth-old girl with surfactant protein B deficiency demonstrating diffuse bilateral granular opacities.

Figure 1B: High-resolution CT axial image of the same patient with surfactant protein B deficiency demonstrating a prominent linear pattern (septal thickening) superimposed on a background of ground-glass opacities (i.e. 'crazy paving').

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FIGURE 2



High-resolution CT axial image in a six-month-old girl with neuroendocrine cell hyperplasia of infancy (NEHI) demonstrating the classic findings of geographic ground-glass opacities seen surrounding the heart and great vessels most conspicuous in the right middle lobe and left lingula.

can provide a very high sensitivity (78% to 100%) and a high specificity (100%) in diagnosing NEHI in the hands of experienced chest radiologists.

Infants affected with PIG are either pre-term or fullterm and usually present within hours of birth and never after six months. Their symptoms include rapid breathing and low oxygen in the blood. PIG has two forms, a diffuse form and a more common patchy form. Imaging findings vary depending on the type of form present. On CT, the diffuse form shows groundglass opacities, thickening of the connective tissue of the lung, and peripheral reticular (spider web like) opacities. In contrast, the patchy form demonstrates multiple small scattered cystic changes of variable size combined with diffuse ground-glass opacities, thickening of the connective tissue of the lung, and peripheral reticular opacities.

Childhood diffuse lung disorders

The final category includes other childhood diffuse lung disorders which typically affect older children. These disorders include multiple different diseases and while this is not a complete list, several of the more common entities and their classic imaging findings are described here.

Langerhans Cell Histiocytosis (LCH)

Langerhans Cell Histiocytosis (LCH) typically affects children from one to 15 years of age and unlike the adult form is unrelated to smoking. Clinical symptoms are nonspecific and 50% of patients with lung involvement do not have any lung symptoms. But LCH in children is usually multi-systemic and rarely just affects the lungs. CT findings demonstrate small nodules which in some cases become cavities. Hypersensitivity pneumonitis

Hypersensitivity pneumonitis is an abnormal inflammatory response to an inhaled allergen. There are three types: acute (4-22 hours), subacute (weeks to months) and chronic. CT findings in acute and subacute cases demonstrate small nodules, ground-glass opacities and air trapping (more lucent areas of the lung) which typically does not affect the upper lungs. Chronic cases demonstrate honeycombing, which is a honeycomb appearance to the lung due to fibrosis of the connective tissue of the lung and cystic changes to the lung tissue.

Cystic fibrosis

Cystic fibrosis is the most common genetic disorder causing chronic lung disease in children and results from a mutation in the cystic fibrosis transmembrane regulator gene. Chest imaging early in the disease may be normal or demonstrate mild air trapping and or dilatation of the airways. CT is more sensitive than even pulmonary function tests in detecting mild or localised lung disease. Later stages demonstrate upper lobe predominate airway dilatation, airway wall thickening, and mucus plugging of the airways.

CONCLUSION

In summary, the diagnosis of childhood ILD has posed significant challenges in the past. However, characteristic imaging appearances of several specific types of chILD are emerging and recognition of these imaging findings has promise for early and accurate diagnosis. Radiology has therefore taken a prominent role in the optimal management of patients with chILD.

FIGURE 3



Figure 3A: Frontal chest radiograph of a two-yearold boy with pulmonary Langerhans Cell Histiocytosis demonstrating bilateral areas of cystic changes.

Figure 3B: HRCT axial image of the same patient demonstrating extensive cystic changes.

FIGURE 5



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HRCT axial image of a three-year-old girl with chronic hypersensitivity pneumonitis demonstrating posterior honeycombing due to fibrosis of the connective tissue of the lung with ground-glass opacities and air trapping (more lucent areas of the lung).



HRCT coronal image in a 17-year-old boy with cystic fibrosis demonstrating dilated airways mainly in the upper lobe, airway wall thickening, and mucus plugging of the airways.



CONGENITAL HEART DISEASE

BY CHARLOTTE DE LANGE

congenital heart defect or anomaly is an abnormality of the structure of the heart and great vessels that is present at birth. It is abbreviated to congenital heart disease (CHD). Heart defects are among the most common birth defects. The incidence varies in different studies, but approximately nine out of 1,000 live born babies are born with some form of heart defect.

There are different types of defects with a variety of complexity. They may either block (obstruct) blood flow in the heart or the great vessels near it, or cause blood to flow through the heart in an abnormal pattern. Some defects are small, of no great concern and without need of treatment, while others are highly complex and may cause serious symptoms that will require medication or surgery, sometimes even immediately after birth.

CHD is frequently diagnosed with ultrasonography while the baby is still in the womb, but re-examination of the baby shortly after birth is needed to confirm the diagnosis. Delayed onset of symptoms and late diagnosis in adolescence or adulthood occur, but are less frequent. With new surgical advances in the last decades, most complex congenital heart malformations can now be treated successfully shortly after birth, and an increasing number of patients survive into adulthood. This has resulted in a growing population of young adults with CHD (ACHD) who need careful monitoring. In recent years there is an increasing general acceptance that a successful treatment for a complex CHD does not lead to a 'normal healthy' heart and careful management frequently includes repeated imaging examinations throughout life.

In this regard, the different radiological techniques used in paediatric radiology play an important role both in establishing the correct diagnosis in the neonatal period, as well as in continuous follow-up throughout life.

THE NORMAL HEART

During early foetal life, there is a complex sequence of developmental stages that results in a normal heart at birth. Disruption of any part of this sequence may result in a defect. There are also physiological changes in the heart and lung circulation during the transition from foetal to neonatal life that may cause disease if they fail to develop.

In a normal heart and circulation, venous (oxygen poor) blood is led towards the heart by two large veins, one from the upper and one from the lower part of the body (the superior vena cava and inferior vena cava) to the right small chamber (the right atrium) and further into the right main chamber (the right ventricle). By a heart muscle contraction, blood from the right ventricle is pumped out to one great vessel (the pulmonary artery) that leads the blood into the lungs, where it is oxygenated. Then arterial (oxygen rich) blood is transported via the pulmonary veins to the left sided small chamber (the left atrium) and then into the left main chamber (the left ventricle), and again by a heart contraction the oxygen rich blood is pumped out to the body through the other great vessel (the aorta). This means that during one heart beat blood is led both into the lungs and out to the body. Between the small and main chambers and between the main chambers and great vessels, there are leaflets (valves) that open and close during contraction to direct the blood flow (Figure 1).



THE MALFORMED HEART -FROM SIMPLE TO EXTREMELY COMPLEX DEFECTS

There is a large spectrum of malformations and combinations of different defects of the heart. Some malformations may result in poor oxygenation of the blood circulation – 'blue baby syndrome'.

Defects can consist of a simple hole in the wall (septum) between the atrial or ventricular heart chambers, called a septal defect, which results in oxygen rich and oxygen poor blood mixing. A hole between the right and left ventricle, called a ventricular septal defect (VSD), is the most common CHD.

However, the heart can be completely changed in its construction and position in the thoracic cage, resulting in changes in size and positions of the ventricles and atria. The position of the great arteries attached to the heart may be inverted, or transposed, (known as 'transposition of the great arteries', resulting in two parallel non-communicating circulations. This is a lethal condition unless a communication between

oxygen rich and oxygen poor blood exists, like a septal defect.

In the complex defect known as a single ventricular situation, there is only a single ventricle working together with a large common atrium and only one of the two great vessels is unobstructed, like in hypoplastic left heart syndrome.

Complex cardiac malformations are often associated with other organ anomalies in the thorax and abdomen, so it is important to diagnose them due to the potential need of treatment.

IMAGING METHODS IN CHD

Cardiovascular imaging of the heart and vessels is a rapidly evolving field that requires familiarity with the appearances of paediatric and adult CHD on chest radiographs as well as images obtained with echocardiography, computed tomography (CT), magnetic resonance imaging (MRI), and cardiac catheterisation.

At the time of the first surgical procedures for complex CHD, cardiac catheterisation was the most important imaging tool for the evaluation of both anatomy and physiology. It remains important for special indications, but from the 1980s, the first line of investigation has been **echocardiography.** The use of two-dimensional grey scale images together with colour and spectral Doppler, will in many cases give an accurate and comprehensive evaluation of the anatomy and function of the heart and the great vessels. It is often sufficient for the evaluation of simple malformations in the neonatal period.

Chest radiography of the heart and thorax can provide important information on the size, shape and position of the heart and great vessels in the thorax, as well as an evaluation of the pulmonary vasculature. Radiographic findings may be suggestive of a heart disease and a specific type of malformation, but are not reliable enough to make a definite diagnosis (Figure 2).

In CHD and ACHD, MRI, CT and cardiac catheterisation are only required to answer specific questions after echocardiography has proved inconclusive. It is important to use individually tailored protocols. In more complex heart malformations and in aortic arch anomalies, imaging with CT or MRI is necessary to show the anatomy and function of the heart. Both methods can display the anatomy of the heart and the great vessels (Figure 3).

In addition, **MRI** can demonstrate the function of the heart with a cine film during the cardiac cycle, showing how the heart muscle contracts and relaxes (Figure 4). If there is narrowing (stenosis) or leakage (regurgitation) due to malfunction of the valves between the atria and ventricles or between the ventricles and the great arteries, it is possible to measure the flow changes as well as calculate the volumes of the ventricles. By using contrast agents, MRI can disFIGURE 1



The normal heart. SVC= superior vena cava, IVC= inferior vena cava, RA= right atrium, RV= right ventricle, PA= pulmonary artery, PV= pulmonary veins, LV= left ventricle, AO= Aorta. Blue arrows= oxygen-poor blood. Red arrows= oxygen-rich blood.

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IGURE 2



Chest x-ray of three newborn babies. A: Normal appearance. B: Right sided heart with round shape in 'Transposition of the great arteries with dextrocardia'. C: Severe enlargement of the heart in a case of Ebstein's anomaly.

play small vessels and structures in more detail and tissue characteristics such as localised and diffuse scarring and inflammation of the heart muscle. MRI can also test the different parameters that can be used to predict patient outcome. Unfortunately, MRI examinations of the heart often take a long time and require general anaesthesia for babies and children under six years of age.

Cardiac CT overcomes many of the drawbacks of MRI. CT is a fast method, but uses ionising radiation to create images of the heart and thoracic cage. Modern CT scanners produce excellent detailed images (better than MRI in small children) and have adapted techniques for radiation dose reduction. For these reasons, and despite the radiation dose, CT is often preferred over MRI in some critically ill neonates and children.

MRI plays its most important role in postoperative evaluation and further surgical planning of CHD and ACHD. In this situation, echocardiography has some shortcomings due to a poor acoustic window through the thoracic wall, partially due to scarring and sternal sutures.

Cardiac catheterisation is an invasive technique that requires general anaesthesia in children and is today reserved for specific haemodynamic evaluation and interventional treatment. By introducing catheters into the vessels and the heart, it is possible to measure the pressure in chambers and vessels, and widen narrow vessels with balloons and close some types of septal wall defects or vessels with plugs. These interventions may replace surgical treatment.

When imaging CHD, especially in young children, the least irradiating and most simple technique that can give a sufficient diagnostic yield should be used, according to the ALARA principle (As Low As Reasonably Achievable).

CHD can be very complex and often require discussion in a multidisciplinary team to make treatment decisions and to decide the further follow-up of patients and their families. Current recommended strategies include serial follow-up with echocardiography, but also cardiac MRI/CT on a regularly basis to evaluate the heart function in children and adolescents/adults with CHD.

In complex CHD, other organs may suffer from the circulatory changes after heart surgery and require investigation and treatment.

In conclusion, remarkable improvements in diagnostics and treatment options have led to increased survival of patients with CHD. Cardiovascular imaging is mandatory in the initial phase in diagnosing the CHD as well as in the lifelong follow-up. CT and MRI have become important adjuncts to echocardiography. CHD should be taken care of in centres with specific expertise in evaluating both the haemodynamic situation and in the interpretation of CT and MRI.

FIGURE



CT angiography A) Axial image showing a right sided aortic arch (short arrow) and B) a volume rendered reconstruction, showing the aorta (in red) with a Kommerell diverticlulum (long arrow) compressing the oesophagus (in yellow) but not the trachea (in blue). The child presented with swallowing difficulties.

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FIGURE 4



MRI cine imaging of the heart showing the four chambers in A) a normal heart, and B) in 'Hypoplastic left heart syndrome' where the left ventricle (LV) is small and the right ventricle (RV) is large.

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PAEDIATRIC LARGE **AIRWAY DISORDERS**

BY MARK C. LISZEWSKI AND EDWARD Y. LEE

INTRODUCTION

A variety of disorders can affect the large airways (trachea and bronchi) of infants and children and many are potentially serious. Congenital lesions, infection, benign and malignant tumours, and aspiration of foreign bodies are the most commonly encountered paediatric large airway disorders in clinical practice. Children are more prone to airway complications than adults because a child's airways are smaller and less stiff, making them more susceptible to narrowing and collapse. Symptoms can vary widely from potentially lethal acute respiratory arrest (stopping breathing) to abnormally noisy breathing, wheezing, recurrent lung infections, and sleep apnoea. Imaging plays an important role in diagnosing these conditions.

IMAGING TECHNIQUES

Radiographs

Plain radiographs are the first imaging test to evaluate the large airways in paediatric patients. Air within the trachea and the bronchi appears black on radiographs, and provides excellent contrast compared to the adja-

cent soft tissues, which appear white and grey. Disorders of the large airway often cause the black column of air within the trachea or bronchi to be narrowed, dilated, obscured or abnormally deviated, alerting the radiologist to the possibility of an abnormality of the airway.

Fluoroscopy

Fluoroscopy is an imaging technique that uses continuous x-rays to capture moving images, essentially a 'movie' version of a radiograph. Fluoroscopy of the airway can be particularly useful because it can show changes in the trachea and bronchi during different phases of breathing. Several disease processes may only be visible during certain phases of the breathing cycle, and may not be seen on standard radiographs. Some of these conditions include floppy airways, including laryngomalacia and tracheomalacia, air trapping in the lungs, and abnormal diaphragm movement¹. A barium swallow examination is often performed along with airway fluoroscopy, during which a patient drinks a barium solution which is radiopaque (visible on radiographs) and images of the oesophagus are obtained. This can be used to assess the patient for additional conditions including abnormal connections between the airway and oesophagus (tracheoesophageal fistulae), ingested food material in the airway, and external structures which may compress the oesophagus and airway.

Computed tomography (CT)

Rapid advances in computed tomography (CT) technology over the past two decades have revolutionised the imaging evaluation of the large airways². Unlike radiographs and fluoroscopy, which produce two-dimensional (2D) images of the entire imaged region, CT produces high-resolution images in cross section, which can provide highly detailed information about the airway and adjacent structures. New advances in imaging technology have allowed CT data to be reconstructed into three-dimensional (3D) images, which produce realistic representations of the trachea, bronchi and distal airways. These 3D CT images can be formatted to create a Virtual Endoscopy (VE), which simulates the appearance of the airways as would be viewed by an endoscopic camera, but without requiring an invasive procedure. CT can be performed with intravenous contrast to highlight the blood vessels of the chest, and abnormal blood vessels that may compress the airway. Similar to fluor-



oscopy, a new CT technique called four-dimensional (4D) CT can acquire multiple CT images throughout the breathing cycle and capture a moving 3D image of the airway. This powerful technique can be used to assess dynamic lesions that may only be visible during certain phases of breathing. CT is a valuable tool in the assessment of large airway disease, but it is always used judiciously because of the potential risks of radiation exposure.

Magnetic resonance imaging (MRI)

MRI is an attractive non-invasive imaging modality, particularly in children, because it uses no ionising radiation and produces high-resolution images in cross section like CT. However, MRI is much more sensitive to motion and other artefacts which can make images blurry and uninterpretable. This poses particular challenges in children who may have difficulty lying still and following breathing instructions (such as "hold your breath"). Recent advances that have made MRI guicker have helped to reduce these artefacts, allowing MRI to be used to look at the large airways³. Sedation is often needed in infants and young children who cannot hold still, but can frequently be avoided in older children who can follow instructions. Like with

CT, intravenous contrast can be given to highlight the blood vessels and investigate vascular lesions that may affect the airway. Cine or Video MRI is a recently developed technology that acquires multiple MR images through the breathing cycle. Similar to fluoroscopy and 4D CT, cine MRI allows the evaluation of dynamic abnormalities of the large airways that may only be visible at certain times in the breathing cycle.

SPECTRUM OF IMAGING FINDINGS

Congenital malformation

Tracheobronchial branching anomalies

Tracheobronchial branching anomalies are caused by a bronchus arising from an abnormal, or ectopic, location. The three main types of ectopic bronchi are tracheal bronchus, cardiac bronchus and oesophageal bronchus⁴. Normally, the trachea divides and forms right and left bronchi, each of which further divides to form smaller bronchi that lead to each lobe of the lung. A tracheal bronchus occurs when an upper lobe bronchus arises directly from the trachea (Figure 1). The incidence is between 0.1-2% on the right and 0.3-1% on the left⁴. A cardiac bronchus is a rarer anomaly where a supernumerary bronchus arises from the right bronchus intermedius. Tracheal and cardiac bronchi usually cause no symptoms, but some children may develop a cough, recurrent infection, collapse of the lung segment supplied by the abnormal bronchus, or stridor (a high-pitched breath sound resulting from turbulent air flow). An oesophageal bronchus is different to tracheal and cardiac bronchi because it arises from the oesophagus rather than another airway. Unlike the other two types of ectopic bronchi, oesophageal bronchi cause symptoms in virtually all patients due to swallowed material entering the lung via this abnormal connection, and are most commonly discovered soon after birth. Tracheal and cardiac bronchi are most commonly detected on CT, while oesophageal bronchus is usually diagnosed via a barium swallow examination.

Congenital tracheal stenosis

The trachea is normally supported by a scaffolding of numerous C-shaped cartilage structures. In rare cases these C-shaped structures can develop abnormally and create a small complete ring leading to congenital tracheal stenosis (Figure 2). Congenital tracheal stenosis is rare but potentially life threatening, with affected paediatric patients typically presenting with breathing problems at birth. Several other conditions are often associated, including pulmonary artery sling, tracheaoesophageal fistula, and pulmonary hypoplasia. Bronchoscopy is the gold standard for evaluation of tracheal stenosis, but CT is playing an increasing role, especially with advances in 3D techniques.

Tracheobronchomalacia

Tracheobronchomalacia (TBM) is a condition in which the airway loses its normal stiffness due to weakening of the cartilage scaffolding that normally supports the tracheal and bronchial walls. Patients can be born with this condition (primary TBM) or it can be acquired as a result of chronic compression from an abnormal adjacent structure compressing the airway (secondary TBM). During inspiration, the airway is often normal in diameter, but when the intrathoracic pressure increases during expiration, the weakened airway collapses. TBM is often underdiagnosed because standard imaging tests performed when the patient has breathed in may not demonstrate any abnormality. The greater use and availability of dynamic imaging modalities including 4D CT and cine MRI have increased detection of this condition^{4, 5}.



(arrow).

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Tracheal bronchus. 3D volume-rendered CT of the lung and airways shows a tracheal bronchus



Congenital tracheal stenosis. 3D volume-rendered CT of the airway and vessels shows focal narrowing of the trachea (arrow).

Vascular rings and slings

Mediastinal vascular anomalies can cause breathing symptoms by compressing an otherwise normal large airway. The most common of these anomalies are termed vascular rings and slings. Vascular rings are vascular anomalies that form a ring around the trachea and oesophagus, often leading to breathing and swallowing symptoms. Vascular rings include double aortic arch, right aortic arch with aberrant left subclavian artery (Figure 3), right aortic arch with mirror image branching and retro-oesophageal ductus arteriosus, and circumflex aortic arch. A pulmonary artery sling occurs when the left pulmonary artery arises abnormally from the right pulmonary artery and passes between the trachea and oesophagus, leading to narrowing of the lower trachea or right bronchus. Untreated chronic tracheobronchial compression from vascular rings and slings can cause secondary TBM, further worsening symptoms⁵. These vascular anomalies may be suspected on radiographs, but can be definitely diagnosed on contrast-enhanced cross-sectional imaging such as CT and MRI. MRI is increasingly replacing CT in the evaluation of vascular anomalies due to its excellent resolution and lack of ionising radiation.

Infection

The three most common infections that affect the paediatric large airways are croup, epiglottitis, and tuberculosis. Croup is typically a mild viral infection that causes upper airway obstruction in children between six months and three years of age. It is most commonly caused by the parainfluenza virus, producing symptoms of fever, inspiratory stridor, and a characteristic barking cough. Imaging is often not necessary to diagnose croup, but radiographs will demonstrate narrowing of the upper trachea, producing a classic inverted V appearance ('steeple sign').

Epiglottitis is a potentially more serious life threatening bacterial infection of the upper airway, which has fortunately become rare with widespread vaccination against Haemophilus influenza type B. Epiglottitis may cause symptoms of fever, sore throat, difficulty breathing and drooling from an inability to swallow. Radiographs show thickening of the epiglottis ('thumb sign'), thickened aryepiglottic folds, and effacement of the vallecullae (Figure 4). Tuberculosis most commonly causes breathing symptoms from infection of the lungs, but airway involvement is another source of breathing difficulty. Lymph nodes can become significantly enlarged in primary tuberculosis infection and may compress the airways. Tuberculosis can directly infect the airway, which leads to necrosis and ulceration of the tracheal and bronchial walls. As the airway heals, it can become fibrotic and narrowed, leading to chronic airway stenosis². Findings of tuberculosis are often first suggested on radiographs, and subsequent CT demonstrates lymphadenopathy and airway complications with much greater detail.

Cancer

In general, cancers are rare causes of large airway obstruction in children. Two potential neoplastic causes of airway narrowing in children are subglottic haemangioma and carcinoid tumour. Subglottic haemangiomas are rare benign vascular tumours. Though benign, subglottic haemangiomas can be life-threatening due to their potential to obstruct the airway². Subglottic haemangiomas may be asymptomatic at birth, but grow in the first weeks of life and cause increasing airway compromise by 1-2 months of age. Due to their vascular nature, they are well seen on contrast-enhanced CT or MRI (Figure 5). If symptomatic, medical or surgical treatment is often needed, but if asymptomatic they are often managed conservatively as most begin to spontaneously regress by one



Vascular ring. A: PA chest radiograph demonstrates a right aortic arch (arrow). B: Axial CT image shows the right aortic arch. C: 3D volume-rendered CT of the airway and vessels from the posterior perspective shows the right aortic arch (arrow) with an aberrant left subclavian artery (arrowhead) encircling the trachea (pink). D: 3D volume-rendered CT of the airway and lungs with vessels subtracted shows narrowing of the trachea at the location of the right aortic arch (arrow).

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year of age. Carcinoid tumours are the most common malignant primary bronchial neoplasm in children². Bronchial carcinoids are tumours of neuroendocrine cells of the bronchial wall which grow into the airway. Affected paediatric patients most commonly have symptoms of wheezing and pneumonia caused by airway obstruction. Radiographs may visualise a mass or nodule, or might detect secondary findings caused by airway obstruction including lung collapse, air trapping, or lung consolidation. These findings are better demonstrated on CT, particularly with intravenous contrast, as carcinoids tend to avidly enhance.

Foreign body aspiration

Foreign body aspiration is a frequent paediatric emergency, with incidence peaking at two years of age. If the aspiration event is not witnessed, diagnosis can be challenging as breathing symptoms may be confused with other conditions. Radiography, fluoroscopy and CT are the three most commonly used imaging modalities in the evaluation of foreign body aspiration. Radiopaque foreign bodies, such as metallic items, are easily seen on any of these modalities. However, most aspirated foreign bodies (90%) are radiolucent, making detection difficult. In these cases, radiographs may show secondary findings of lung hyperinflation or collapse, but they are frequently normal. Therefore, CT is being used with increased frequency. CT may be able to visualise a foreign body not seen on radiographs, and has a superior ability to detect secondary findings of lung hyperinflation and collapse. The addition of 3D and 4D CT techniques has also increased the sensitivity², however bronchoscopy remains the gold standard for the detection of aspirated foreign bodies.

CONCLUSION

Imaging plays an essential role in the evaluation of various congenital and acquired large airway disorders in children. Radiographs are often the first imaging test and are sometimes valuable in detecting large abnormalities of the airway. CT and MRI are able to show structure in great detail and have become essential tools in the diagnosis of many of these conditions, particularly when radiographs are unhelpful. New and exciting 3D and 4D imaging techniques have extended the diagnostic capabilities of imaging, furthering the ability to diagnose paediatric large airway disorders without the need for invasive testing.



Epiglottitis. Lateral radiograph of the neck demonstrates thickening of the epiglottis (arrow) and thickened aryepiglottic folds (arrowhead) (Case courtesy of Terry Levin, MD).

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Subglottic haemangioma. Coronal image from T1-weighted MRI of the neck demonstrates a subglottic mass (arrow) causing airway narrowing (Case courtesy of Terry Levin, MD).

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IMAGING TUBERCULOSIS

BY BERNARD F. LAYA AND KHRISTINE PULIDO-BRILLO

GLOBAL TUBERCULOSIS BURDEN

Tuberculosis (TB) is an airborne infectious disease of global public health concern with one-third of the world's population being infected. Its burden is enormous and it ranks as the second leading cause of death from a single infectious agent, after human immunodeficiency virus (HIV). TB is present in all regions of the world but most cases are in south-east Asia, Africa and the western Pacific. In 2013 alone, an estimated nine million people developed TB (13% with HIV co-infection) and 1.5 million died from the disease. The same year, an estimated 550,000 children became ill and 80,000 died from TB. There is also a worsening problem of multi-drug resistant tuberculosis (MDR-TB), which is seen in patients with TB that does not respond to the usual TB medication currently available. Access to TB care has expanded through the years, which has helped to reduce TB mortality by 45% since 1990, and millions of lives are being saved through effective diagnosis and treatment. The global TB prevalence continues to improve with its incidence declining at an average of 1.5% per year between 2000 and 2013. Despite this, mortality from this preventable and curable disease remains unacceptably high. TB is not simply an infectious disease but also a reflection of overall social conditions, commonly affecting poorer countries.

TUBERCULOSIS INFECTION AND DISEASE PROGRESSION

TB is a transmittable disease caused by infection with the *Mycobacterium tuberculosis* bacillus. It typically affects the lungs (pulmonary) but can affect other organs of the body. It is spread when people with pulmonary TB disease expel the organisms primarily by coughing. Once inhaled, the mycobacteria organisms settle in the lungs and lymph nodes causing a series of inflammatory reactions. If a child is healthy, the infection is contained but not totally eradicated, and the bacilli become dormant. This condition is referred to as latent tuberculosis infection (LTBI) where a person tests positive to a TB skin test or other TB laboratory tests, but may not have any physical manifestations of TB disease. Children are infected through exposure to infected adults and contribute little to disease transmission. Of infected children, 95% develop the latent infection and will not develop the actual TB disease. In individuals with a poor immune state, including malnutrition, immune suppression (including HIV infection), and young age (less than five years old), the latent infection progresses into active TB disease. Progression of the disease manifests in the lungs and lymph nodes, as well as other structures in the chest and distant body parts as the infection reaches the blood stream. Poverty and poor living conditions (resulting in malnutrition and crowding), HIV co-infection, and lack of appropriate anti-TB drugs are the most significant causes of TB disease progression in many developing countries.

SIGNS AND SYMPTOMS

Clinical signs and symptoms can be diverse and could depend on the location and extent of disease, as well as age and immune status. TB can affect every organ in the body, but pulmonary infection is by far the most common. Children with pulmonary TB may have no symptoms, or may present with nonspecific symp-



toms such as a longstanding cough, fever, palpable lymph nodes, loss of appetite, and weight loss, which are hard to differentiate from other causes. If TB is left untreated, symptoms and signs of severe lung disease and other organ involvement maybe apparent.

DIAGNOSIS AND MEDICAL IMAGING OF TB

TB in children remains a diagnostic challenge. Identification of the TB bacilli in sputum seen under a microscope and TB culture are definitive tests in adults but are not routinely attempted in children because they do not produce enough sputum for the test. A positive TB skin test suggests infection but cannot differentiate active or quiescent disease, and a negative TB skin test does not exclude TB. A highly sensitive molecular laboratory test (Xpert MTB/RIS) is now available but still, a negative result does not exclude TB. History of TB exposure, signs and symptoms, laboratory and microbiologic tests are all important diagnostic tools but medical imaging tests also remain valuable in TB diagnosis. In
some instances, imaging offers the only way to thoroughly evaluate the extent of the disease.

Medical imaging methods are important in screening, diagnosis and thorough evaluation of TB, which have important treatment implications. It helps to assess complications, detect other underlying diseases, including HIV, and helps in monitoring during and after administration of treatment. The chest radiograph (x-ray) is widely used because of its availability and instantaneous results. A normal chest x-ray cannot exclude the presence of pulmonary TB, but when positive, the findings closely reflect the extent of the disease. Ultrasound is easy, inexpensive, non-invasive, and useful for identifying abnormally enlarged lymph nodes and abnormal fluid collections in the chest. Computed tomography (CT) is the examination of choice in unusual, complicated, and extensive disease. The advantage of CT over chest x-ray is in defining the extent of the disease and its possible complications within and outside the chest. MRI is also an excellent method for detecting chest lymph nodes, but provides poor detail in visualising the lungs.

The presence of enlarged lymph nodes in the chest, with or without a visible lung abnormality, is the most common x-ray feature of childhood TB infection, seen in 92% of cases (Figure 1). Even with adequate treatment, these lymph nodes may calcify and may persist for months and even years. On chest x-ray, these manifest as rounded structures, which may cause compression of the airway. If TB disease progresses, the lymph nodes enlarge and develop cavities that can compress the airway and block airflow to the lungs.

The primary focus of infection in the lungs may be too small to be visible on the chest x-ray but it commonly appears as dense, patchy or linear densities. Cavitation or calcifications can be seen within the lung and can progress to extensive lung damage (Figure 2). TB can reach the lining of the lungs and the heart and can lead to an accumulation of thick abnormal fluid collections that can compromise both the lung and heart functions. Although large fluid collections can be seen on chest x-ray, there is improved visualisation and more accurate measurement of the fluid with ultrasound, MRI or CT imaging. The infection can also reach the bloodstream and affect virtually any organ in the body including the brain, liver, kidneys, intestinal tract and bones. Findings of numerous, fine nodular opacities throughout the lungs, along with findings of TB meningitis, are indications of disease spread in the bloodstream.

TB that was initially latent or dormant can reactivate especially in older children and adults with declining health and immune status. On imaging, reactivation appears as ill-defined lung densities mainly involving the upper lobes of the lungs. It is usually associated with nodular and linear scarring, cavities and eventual destruction of portions of the lung (Figure 3). Radiographic and clinical evaluations also become major indicators of response to therapy. Regression is a slow process and chest x-ray findings may get worse before they get better. Resolution of lung abnormalities has



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Nine-year-old boy with primary tuberculosis. Frontal chest x-ray (A) reveals a soft tissue fullness in the right perihilar area of the chest, indicative of enlarged lymph nodes (black arrow). CT scan of the chest in transverse view (B) confirms the enlarged lymph nodes (white arrow), typically seen in primary tuberculosis.





Primary progressive tuberculosis in a six-month-old infant presenting with fever, difficulty breathing and positive TB skin test. Frontal chest x-ray (A) shows right upper lobe consolidation with suspected cavity formation (black arrow). CT scan of the chest in transverse view (B) confirms the right lung disease with cavities (white arrow), and enlarged lymph nodes in the chest (asterisk).



A 17-year-old boy with reactivation of tuberculosis presenting with fever, cough, and blood-tinged sputum. Frontal chest x-ray shows a left upper lung disease with fibrotic strands, cavities, and volume loss (arrow), all of which are indications of reactivation of a previously dormant tuberculosis infection.

TREATMENT AND OUTCOMES

References

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been observed from six months to two years on chest x-ray and up to 15 months on CT scans. Lymphadenopathy may persist for several years after treatment.

Without treatment, the TB death rates are as high as 70% within ten years. Anti-mycobacterial therapy is the cornerstone of TB treatment, taken for at least six to nine months depending on age, overall health, possible drug resistance, organ involvement, and form of disease. Children with latent infection should be treated because of the high risk of disease progression, a longer remaining potential lifespan in which the disease can develop into active disease, and the potential for spreading the disease. TB infection that has reached the bloodstream and other body parts requires longer and more aggressive treatment. Early initiation of antiretroviral therapy (ART) and isoniazid preventive therapy (IPT) has improved treatment outcomes in patients with TB-HIV co-infection. Directly observed therapy programmes, where a healthcare worker helps to administer the medication, have helped to increase medication intake compliance. Completing the treatment course is essential to avoid the development of drug resistant strains that are much more dangerous and difficult to treat. In 2013, the treatment success rate continued to be high, at 86% among all new TB cases. Treatment for multidrug-resistant TB is longer, and requires more expensive and more toxic drugs.

CONCLUSION

Diagnosis of pulmonary TB is a continuing challenge to both clinicians and radiologists. Understanding the cause, complications, and the global impact of the disease is important. Careful clinical history, TB skin testing, laboratory testing, and chest x-ray remain the basic elements for establishing diagnosis. Other imaging tests, including ultrasound, MRI, and CT, can provide relevant and more detailed imaging information about the disease. Bringing together public health agencies, the pharmaceutical industry and academics can defeat TB, but since TB is a global healthcare concern, public awareness about the disease is very important.

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IMAGING THE ABDOMEN

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RENAL TRACT IMAGING

BY LIL-SOFIE ORDING MULLER

INTRODUCTION

Disorders of the renal tract are one of the most common reasons for imaging in childhood. The range of diseases is wide, from urinary tract infection, which affects up to 8% of all children, to complex malformations and renal failure. Imaging plays a key role in the diagnosis and management of children with disorders of the renal tract.

BACKGROUND

The normal renal tract consists of two kidneys which filter the blood to make urine, the collecting system (ureters), which directs the urine to the bladder, and the urethra, through which the bladder empties. In a normal child, the two kidneys work equally well. The kidneys not only produce urine and regulate body electrolytes, but also produce a variety of hormones, including those involved in blood-pressure regulation.

On urination, the bladder contracts, the muscle at the base of the bladder opens and the urine flows through the urethra. In a normal system, the urine passes through the ureters and into the bladder without a problem. The angle between the bladder wall and the ureters normally gives a 'valve-effect' preventing backward flow of urine from the bladder. One of the most important abnormalities is the backward flow of urine up the ureter to the kidney. This is known as vesico-ureteric reflux (VUR), and is common in young children. Whilst mild cases of VUR may improve as the child grows, and treatment may only be required for urine infections if they occur, many children will not grow out of more severe disease.

The kidneys, collecting system, bladder, urethra and their blood vessels can be thought of as working together as one unit, hence why we use the term 'renal tract' for this system. Abnormalities in one part of the system may affect the others, for instance, blocking after birth.

IMAGING MODAL ITIES

Ultrasound (US) is the main imaging technique for the paediatric urinary tract. Children have relatively little body fat, which makes ultrasound easier. New ultrasound machines with high-resolution probes allow highly detailed images of the whole urinary tract including the kidneys (Figure 1). US is an easily accessible tool with no ionising radiation, meaning it



urine flow in the collecting system may cause high backward pressure, which subsequently may damage the kidneys. Congenital abnormalities of the kidneys and urinary tract account for 30% of abnormalities seen in children, and can cause the kidneys to fail to work (renal failure). It is therefore important for those who image children to have a good knowledge of the basic development of the kidneys in foetal life and the normal age-related appearance of the urinary tract

is ideal for use in children. It allows both anatomical and dynamic assessment and can also be used to assess flow in blood vessels.

The US scans should ideally be performed by an investigator experienced in paediatric radiology, or be supervised by a paediatric radiologist, using an up-to-date method for paediatric imaging.

Fluoroscopy

In fluoroscopic examination of the renal tract, contrast (dye) is usually placed into the bladder via a catheter and the child is examined during urination or 'voiding', an examination called voiding cysto-urography (VCUG). This allows assessment of bladder contraction and emptying, as well as the bladder wall and the urethra. During filling of the bladder and urination the paediatric radiologist also looks for reflux of contrast from the bladder back into the ureters (VUR; Figure 2), as well as blockages in the urethra

Nuclear medicine studies

Unfortunately, although ultrasound provides excellent anatomical information about the kidneys it cannot currently tell how well they work. In nuclear medicine studies, a small amount of radioactive material is injected into the blood and excreted through the kidneys. Some radioactive tracers can show whether the kidneys are scarred, or whether there is ongoing infection of the renal parenchyma (e.g. using Tc-99-m DMSA), whereas others which pass through the kidneys can show how well they are working (Tc-99m MAG3). MAG3 can therefore tell whether one kidney is working better than the other (Figure 3). Both MAG3 and DMSA can be used to calculate the split renal function i.e. the percentage contribution of each kidney to the total renal function.

MRI and CT

MRI can give both detailed images and tell us about the function of the kidneys, without using ionising radiation. However, usually MRI scans of the kidneys take a long time, often over an hour, and therefore may need sedation in young children. It is particularly useful in complex malformations of the kidneys, and as an alternative to nuclear medicine studies to check kidney function.

CT is still the quickest and best way of examining the kidneys for severe injury during trauma, or to look for dense stones in the kidneys. Otherwise CT is rarely used in imaging the renal tract in children due to the relatively low tissue resolution in children with little body fat, but most of all because of the ionising radiation.

FREQUENT CONDITIONS REFERRED FOR IMAGING

Urinary tract infection (UTI)

Imaging in UTI is performed depending on age and clinical presentation of the infection. The aim of imaging in UTI is to detect underlying conditions such as congenital malformations or VUR, which may mean that the child is more likely to get renal damage, but also to detect voiding abnormalities. Renal damage may cause reduced function or high blood pressure. The main modality in UTI is US, with nuclear medicine tests and MRI used in more complex cases. Follow-up scans are used to check the kidneys' growth and development.

Prenatal abnormalities

Many abnormalities of the renal tract are detected before the baby is born, during pregnancy. Antenatal ultrasound is becoming particularly useful for detecting babies at risk of kidney problems, and indicating what treatment they may need when they are born. This can be particularly useful to prevent the baby from getting any symptoms (e.g. Figure 4).

Dilatation of the collecting system

Expansion or dilatation of the collecting system is often referred to as 'hydronephrosis' (water on the kidney)



Ultrasound scan of a three-year old girl with recurrent urinary tract infections. Ultrasound scan shows a normal right kidney (A) and a small left kidney (B) with dilatation of the collecting system (arrowheads). In the bladder there is urine with hyperechoic spots, which can be seen during a urinary tract infection. Behind the bladder there is a dilated left ureter (C, black arrow). This girl had known left-sided vesico-uretral reflux (see also figures 2 and 3).



Voiding cysto-urography (VCUG) performed in the same patient as in figure 1 shows reflux of contrast from the bladder (arrow) and into the left ureter and renal pelvis (arrowheads).

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A Tc-99m DMSA-scintigraphy of the same patient as shown in figures 1 and 2 shows parenchymal loss of the left kidney with reduced function compared to the right side. The left kidney was calculated to contribute 18%, and right kidney 82%, of renal function (% total relative uptake).





Ultrasound scan of a newborn girl with prenatally detected severe hydronephrosis on the right side. An ultrasound scan after birth showed a normal left kidney (A) and severe dilatation of the renal pelvis on the right side and thin, hyperechoic parenchyma (B-E). Doppler examination showed circulation of the parenchyma (C). There was no dilatation of the ureter. Cortical cysts can be seen using a high frequency ultrasound probe (D, E). These images suggest damage to the renal parenchyma due to an obstruction of the junction between the left renal pelvis and ureter (uretro-pelvine junction obstruction).

Renal cystic disease

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and 'hydroureter' (water in the ureter) depending on the level of the problem. Expansion of the ureters, for example, can occur with VUR, with blockage, or because there is a congenital abnormality in the way the tract was made. Imaging is useful for diagnosing these conditions, assessing how the kidneys function, and indicating what further treatment may be needed.

Incomplete bladder emptying can lead to UTI but also a potential high backward pressure to the kidney. Both may subsequently cause damage to the kidneys. Imaging is useful for determining the severity, level and ideally the cause of the problem.

Cysts in kidneys are relatively common in adults, but quite rare in children, and usually indicate an underlying condition. Cysts may occur in one or both kidneys, and may be identified at any age. Cysts in kidneys may be associated with cysts in other organs, which can usually be checked using ultrasound. Sometimes kidneys do not form properly and appear to be a bag of cysts at birth, which usually do not function properly.

SUMMARY

Imaging plays a major role in the diagnosis and follow-up of both mild and complex diseases of the renal tract. US is the main modality used in children but other advanced methods, like MRI and nuclear medicine studies, may be used together to give both anatomical and functional information.



LIVER IMAGING

BY STÉPHANIE FRANCHI-ABELLA

he liver is an important organ found in the upper right part of the abdomen, below the diaphragm. It is made up of a complex network of specific liver cells (called hepatocytes), bile ducts (which drain the bile to the bowel for digestion) and blood vessels (portal vein, hepatic artery and hepatic veins).

Working together with the kidneys, the liver plays a major role in getting rid of the body's toxins from the blood, acting as a filter. In particular, the liver filters the blood coming from the bowel, which contains all the food absorbed during digestion. Only after this step of detoxification can the blood flow into the rest of the body. Aside from detoxifying the blood, the liver has several other important functions including producing proteins such as albumin or blood clotting factors, and excreting substances that contribute to digestion (found in bile).

There are many causes of liver diseases in children, including infection, toxic and genetic disorders, tumours, and malformations. Disorders may be transient (e.g. viral hepatitis) or long term (e.g. cystic fibrosis). They may preferentially affect liver cells, bile ducts or vessels.

The severity of liver disease is related to several features: first, the ability of liver cells to continue to produce proteins; second their ability to filter the blood coming from the bowel; and third, the presence of fibrosis in the liver that corresponds to scarring after an injury. When protein production is compromised to the point that there are not enough vital proteins being made for the body to function, then this is termed 'liver failure' with a high risk of death. Currently, there is no treatment that can replace the function of the liver and liver transplantation may be the only therapeutic option.

The inability to filter the blood coming from the bowel may lead to several complications, mainly neurological impairment (hepatic encephalopathy) and cardiovascular complications. In the case of fibrosis, liver stiffness is increased and this sometimes impedes the passage of blood coming from the bowel through the liver. This phenomenon leads to an increase of pressure in the portal veins, (called portal hypertension) and can give rise to heavy bleeding from the digestive tract that can be life threatening. In some severe and rare cases of portal hypertension, the hepatic veins are also involved.

The diagnosis of liver disease relies on the combination of clinical tests, biological markers, imaging and pathological data. Liver imaging plays a vital role not only in the diagnosis and anatomical assessment but also in treatment in certain cases. FIGURE 1



FIGURE 2



This two-month-old girl presented with acute fever and jaundice. Ultrasound showed that the bile duct was enlarged because of stones (white arrow) (A). Using interventional radiology techniques the stones were pushed through into the bowel using the injection of contrast medium and inflatable balloons, the obstruction was relieved as shown by the contrast flowing through the bile duct into the bowel (B and C).



Twenty-two month old baby presenting with a rapid enlargement of his belly. CT scan of the abdomen after injection of intravenous iodine shows a large heterogeneous mass arising from the liver. CT identifies the precise location of the lesion in order to select the best treatment for the patient. This large tumour, called a hepatoblastoma, was treated with chemotherapy and surgical resection.



The first step in the investigation of liver disease is usually ultrasound (US) with Doppler studies. Doppler is an ultrasound technique which is used to demonstrate flowing blood. Indeed, the qualities of US (wide availability, absence of radiation and low cost) mean that it is extremely useful in the diagnoses of hepato-biliary diseases. Moreover, US is an excellent tool in paediatrics because the relative lack of fat in children allows excellent image quality. US with Doppler provides information on the liver, the vessels, and the bile ducts. In addition, US can be used to guide biopsy of a specific lesion within the liver.

In some cases, other complementary techniques such as computed tomography (CT) and magnetic resonance imaging (MRI) are necessary to make the diagnosis. These will provide images of the whole abdomen and may require the injection of intravenous contrast media (lodine for CT and Gadolinium for MRI). The images obtained can be reconstructed in different ways to show different views of the liver and its components and can give more precise information about the anatomy and the characterisation of the disorder.

A recent new ultrasound application called elastography, allows the measurement of liver stiffness, which shows promise in the assessment of liver fibrosis and may, in the future, mean we can avoid liver biopsy in some patients.

Interventional radiology uses either ultrasound or x-rays (angiography or CT) to guide more invasive procedures involving the placement of catheters in

vessels, bile ducts, the gallbladder or the liver itself. At present, thanks to major technical advances, the diagnosis is usually made with non-invasive tools, such as US, CT or MRI, while the use of interventional radiology is limited to treatment guided by imaging.

Imaging will sometimes identify the precise diagnosis (e.g. gallstones, obstruction of a vessel, liver tumour); it will show if the liver displays signs of chronic disease (the end stage being cirrhosis), and it can show complications related to the liver disorder such as portal hypertension with increased spleen size and the presence of enlarged vessels around the digestive tract. Elastography will often play a role in the assessment of disease severity without biopsy.

When imaging and blood tests cannot provide the precise diagnosis, it will sometimes be necessary to get a specimen of the liver (biopsy) for microscopic analysis by the pathologist. A liver biopsy involves taking a little piece of the organ using a needle. The procedure is performed using image guidance (ultrasound) to increase the safety of the procedure and if necessary to enable a particular region or area of abnormality to be sampled.

Interventional abdominal imaging can be used in the treatment of some disorders. Being much less invasive than classical surgical access it can replace surgery in some instances. For example, if there is a narrowing of a vessel or a bile duct leading to dysfunction of the liver, it is possible to get into the bile duct or the vessel, using a needle through the skin and a catheter threaded through it. The stricture can be treated by



MRI of the liver using a specific programme to show the bile duct without any injection of contrast media. This eight-year-old girl had a tumour removed when she was two years old. MRI perfectly shows a narrowing of the bile ducts (so-called stenosis, large arrow) and dilatation of the bile ducts above the strictures (thin arrow) which are a side effect of the surgery. Only the injection of contrast media directly into the bile ducts as in figure 2 could provide similar information.

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inflating a balloon and, if necessary, implanting specific devices like stents to keep the vessel open and patent. In some cases, the problem is the presence of abnormal vessels that need to be obstructed. Once again interventional radiology allows us to directly inject glue or special coils into the vessels and occlude the abnormal vascular structures.

When the liver function is really poor, the only remaining therapy may be liver transplantation. Imaging also plays a major role here: first, before the transplantation, in planning the surgery by providing a precise visualisation of the patient's anatomy, particularly with regard to the vessels. Second, during the transplantation, in assessing the patency of the vascular anastomosis (connections between the graft's vessels and the patient's vessels), and third, after the transplantation, in detecting complications in the short, medium

and long term. Follow-up after liver transplantation uses US as the first tool. Once again, interventional radiology plays a major role in the treatment of some of the complications, mainly vascular and biliary. Imaging guidance of liver graft biopsies is mandatory, as the graft is frequently only a part of a larger liver in children.

As described above, paediatric radiologists have an important role in the management of liver diseases in children, providing important information for the diagnosis and the detection of complications at the time of diagnosis and during follow-up. It is less commonly appreciated that they also participate in the treatment of some patients using minimally invasive interventional radiology techniques and helping to avoid the need for invasive surgery.

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Stéphanie Franchi-Abella



PELVIC IMAGING

BY LAURENT GAREL

he pelvis is the lower part of the abdominal cavity that projects below the 'belly-button'. It contains part of the bowel (e.g. the rectum), the bladder, the uterus and the ovaries.

There are several types of pelvic abnormality in children, such as pelvic pain, urinary infection, problems with advanced or delayed puberty, and pelvic masses or cancers.

MAIN INDICATIONS OF PELVIC IMAGING IN CHILDREN

Imaging may focus on a precise anatomical area, either urinary, genital, or digestive, or it may be complex in location and nature.

Nowadays, foetal ultrasound is routine in pregnancy management. Several known abnormalities mean that imaging is carried out before and after birth, even when the conditions are asymptomatic.

Imaging is also important in the assessment of pelvic pain, a frequent symptom in children. Pain during passing urine or stools can be addressed by targeted procedures of investigation.

Imaging is rapidly indicated in cases where a pelvic mass is discovered by the parent or doctor during a medical examination.

Imaging, especially ultrasound, is important for assessing the hormonal status of girls in regard to puberty.

Pelvic imaging is used in the investigation of urinary tract infection, especially when associated with fever; in the search for ectopic testes (i.e. testis not felt in the normal place in the scrotum); in the work-up of significant constipation; and also in the surveillance of some children with previously treated disorders (e.g. follow-up of an ovarian tumour) or those who are genetically prone to developing a familial disorder.

PELVIC IMAGING TECHNIQUES IN INFANTS AND CHILDREN

When imaging children, safety is crucial. Infants and children are more sensitive to ionising radiation than adults. In addition, the location of the ovaries and testes means that pelvic examinations must be performed especially carefully with regard to radiation protection.

Pelvic ultrasound

Ultrasound waves are not transmitted through air, so warm gel is applied to the patient's skin to aid transmission. A full bladder is also important, as it moves the bowel (which contains air) out of the pelvis and into the abdomen. Drinking prior to the examination is therefore useful. Fortunately, children are small and relatively thin compared with adults, allowing the use of high resolution ultrasound equipment. From an ultrasound standpoint, 'small is beautiful', but small is also impatient. Thankfully, ultrasound is fast, and is performed in the presence of parents, without sedation or restraint.

Magnetic resonance imaging (MRI)

Magnetic resonance imaging (MRI) is more demanding than ultrasound, because of the duration (20 to 45 minutes) of the examination, requiring the use of sedation or anaesthesia under the age of six to eight years. MRI is always performed following an initial ultrasound. MRI is based on the magnetic properties of certain atomic nuclei (e.g. the hydrogen nucleus



Ultrasound (sonography) is the most important technique in childhood pelvic imaging. Because of its simplicity, wide availability, speed and reliability, sonography is the first and often the only imaging modality needed when investigating the paediatric pelvis.

present in water molecules, and therefore in all body tissues). Like ultrasound. MRI does not use ionising radiation, which is particularly useful when imaging the pelvis.

When MRI is used to image the urogenital system it is called MRU (MR urography) and when used to image the digestive system it is called MRE (MR enterography).

MRU can be used to display complex anomalies of the urinary tract and can also help to assess renal function and obstruction after intravenous injection of pharmaceutical agents.

MR enterography, where a volume of liquid swallowed by the patient is tracked through the bowel using MRI, is mainly used for the diagnosis, staging and follow-up of inflammatory bowel disease (IBD). IBD is more and more prevalent in children and teenagers and MRE has replaced CT in imaging these patients.

Contrast studies: the impact of modern digital fluoroscopy

Technological improvements have substantially decreased the dose of medical radiation, thanks to digital fluoroscopy, video capture and low-dose CT.

Fluoroscopy is a technique that uses radiation to examine organs in real time. In the pelvis, it may be used to perform cystourethrography where the bladder is observed after the injection of contrast (radiopaque dye) into it. Similarly, the rectum and large bowel can be examined during a barium enema. New equipment delivers the x-rays for fluoroscopy in bursts, leaving short gaps between them (pulsed fluoroscopy) and images are now 'captured' from the screen rather than requiring separate exposure (video capture). This means the traditional contrast studies of the pelvis, cystourethrography and barium enema, have become extremely low-dose examinations. Nowadays, the radiation dose from paediatric cystourethrography can average as low as the equivalent of only ten days of background radiation.

Cystography still requires a catheter to be inserted into the child's bladder and a barium enema requires the placement of a rectal tube, in order to fill the urinary or digestive system with contrast. Obviously, the expertise, equipment and a favourable environment, in a paediatric setting where parents are strongly encouraged to attend the examination with their child, is helpful. In most paediatric institutions, explanatory leaflets are given to the parents in advance, to prepare the child for the procedures and decrease the amount of stress at the time of examination.

Bladder opacification shows and grades the abnormal back flow of bladder contents toward the kidney, which is termed vesicoureteral reflux (VUR), as well as displaying the urethra (tube through which the bladder empties) during voiding sequences in patients with urinary tract infection or urinary obstruction.

Retrograde opacification of the rectum and colon is used to investigate cases of intestinal obstruction, narrowing of the bowel, severe constipation or after surgery.

Pelvic CT

Nowadays, CT is exclusively performed in paediatric patients with tumours, at presentation and during follow-up, examinations are governed by national or international protocols. Technical parameters are always set according to paediatric standards, in order to minimise the radiation.

PELVIC IMAGING GUIDELINES IN CHILDREN

Pelvic masses

Feeling a pelvic mass in a child is an urgent indication for imaging. Ultrasound proves extremely valuable in these circumstances, showing an enlarged obstructed bladder, the presence of impacted stools, a distended, obstructed vagina, or another space-occupying lesion. Ultrasound will show the location of the mass, its cystic or solid nature, and its blood supply, thanks to colour Doppler ultrasound. Subsequent investigations, especially MRI, can be planned subsequently using the ultrasound images for guidance.

Urinary tract infection (UTI)

In most cases of UTI with fever, ultrasound and cystourethrography prove both useful and complementary.

In 2011, the American Academy of Pediatrics updated its guidelines and recommendations relative to UTIs in infants and children with a fever. Ultrasound is indicated in the assessment of the first UTI, whereas cystography is proposed in cases of recurrent infection. Renal scintigraphy (nuclear medicine) or functional MRU can be useful in selected cases for assessing kidney function, scars and obstruction of the urinary tract.

FIGURE 1



11-month-old female with premature thelarche (breast budding). Ultrasound of the pelvis showing a normal infantile uterus (i.e. without sign of hormonal stimulation). The anterior-posterior diameter (in-between calipers) does not exceed 10 mm.

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FIGURE 2



8-month-old investigated for urinary tract infection. Cystography (oblique view) displaying bilateral vesico-ureteral reflux, grade 2/5 along a single left kidney, grade 3/5 along a duplex (double) right kidney. The two right ureters merge into a single conduit prior to entering the bladder (arrow).





3-day-old male infant with prenatally recognised hydronephrosis (dilated kidneys) associated with an enlarged bladder. Voiding cystourethrography (oblique view). The bladder wall is irregular, due to the presence of a congenital urethral obstruction (posterior urethral valves), well-seen on the opacified urethra (arrows). The valves were resected endoscopically.



13-year-old female with cyclic pelvic pain and palpation of a midline pelvic mass. A: Longitudinal ultrasound scan. The uterus is markedly enlarged with menses retention, while the cervix appears non-patent. B: Sagittal MR scan confirming the haematometra (blood retention within the uterus) with uterine cervix aplasia. The patient underwent a hysterectomy

Pelvic pain

Pelvic ultrasound and pubertal status in girls

Some young girls develop signs of puberty very early, often this is not of any concern but sometimes it is prompted by a major hormone imbalance, which requires treatment. Ultrasound can help to tell if this

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Ultrasound is invaluable as the initial imaging modality to search for appendicitis, IBD, ovarian cysts or ovarian torsion (twisted ovary), urinary stones or urinary dilatation, or vaginal obstruction.

Postnatal investigation of a foetal pelvic abnormality

Ultrasound is again favoured as the main tool to show the anatomy. In 2% to 5% of antenatal ultrasound examinations worldwide there is some evidence of fluid accumulation within the foetal kidney, post-natal ultrasound is used to clarify the situation and to gauge whether follow up or further imaging is required. Depending on the complexity of any abnormality, MRU and fluoroscopy can prove useful, especially if surgery is being considered.

is the case, based upon the features and measurements of the uterus and ovaries.

In adolescents presenting with delays in starting menstruation, uterine and ovarian sonographic characteristics are key to working out where the problem lies.

Pelvic ultrasound for ectopic testes

In case where the testis is not present within the scrotum, ultrasound will search for and locate the gland along the inguinal canal in most instances. Surgical consultation is required for these patients, to identify the need for and timing of surgery.

CONCLUSION

In conclusion, ultrasound is the starting point for imaging the pelvic contents in children. This helps to guide a management plan and subsequent investigations, always taking into account the value, the hazards and the discomfort of every procedure.

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PAEDIATRIC CANCER IMAGING

Y TUMOURS (NEUROBLASTOMA AND WILMS TUMOUR) GING IN PAEDIATRIC LYMPHOMA



BODY TUMOURS UROBLASTOMA $\supset M/II MS$

BY ANNEMIEKE LITTOOIJ AND BETH MCCARVILLE

INTRODUCTION

Neuroblastoma is the most common tumour arising in the body (outside the brain) in young children. The clinical presentation varies with the size, spread and location of the tumour, often in an unwell child. The combination of nuclear medicine investigations, where a small amount of a radioactive substance is injected into the body and its distribution in the body is detected with a special camera known as a gamma camera, and radiological studies (x-rays, CT and MRI) are important to assess the spread of the disease and for follow-up.

Wilms tumour is the most common tumour of the kidney in children. Wilms tumour typically presents as a lump in the abdomen in an otherwise well child. Imaging plays an important role in diagnosis and follow-up of the disease

In this chapter, the clinical aspects and imaging features of both tumour groups are briefly described.

NEUROBI ASTOMA

Background

Neuroblastoma (NBL) is a tumour of nerve tissue that forms from immature cells of the sympathetic nerve system. This part of the nerve system controls body functions such as heart rate, blood pressure and the digestive system. NBL develops in infants and children and can occur in many areas of the body, usually in the abdomen, in the tissues of the adrenal gland. Cancer spread is called metastasis, and NBL can metastasise to other areas of the body, including the lymph nodes, liver, bone and bone marrow. In nearly 70% of children with NBL it will have already spread by the time the condition is diagnosed. NBLs have a variable course. Some tumours behave aggressively, while others, typically in infancy, may spontaneously regress. The extent of spread, patient age and the subtype of the tumour are all known to be factors associated with this variable course.

Clinical features

Imaging

surgery.

Magnetic resonance imaging (MRI) is preferred to computed tomography (CT) for imaging NBL in children, because MRI does not use radiation but provides excellent images of the soft tissues. Importantly, MRI is superior to CT in assessing metastatic bone marrow disease, chest wall invasion and spinal canal involvement. MRI can be used to outline the tumour's borders, confirm its size, location and spread, and assess evidence of internal bleeding and calcifications. Despite their size and aggressive behaviour, NBLs tend to wrap around and push adjacent structures out of the way, rather than invade them (Figure 1).

An important feature of malignant tumours is the accumulation of cells which are packed together more closely than in normal tissue. This can be assessed with diffusion-weighted imaging (DWI), a specific sort of MRI scan. Diffusion weighted imaging measures the random motion of water protons. This motion becomes less in parts of the body where there

Paediatric Imaging

The way that NBL is discovered varies with the size and location of the tumour. In the abdomen it can cause abdominal enlargement, sometimes nausea or pain. Unexplained fevers, bone pain or limping are caused by bone or bone marrow involvement. If the tumour is compressing the spinal cord it causes muscle weakness or problems with urinating. Some tumours produce substances that can cause high blood pressure, increased heart rate, facial flushing or diarrhoea.

Ultrasound is often the first investigation performed in children presenting with abdominal disease. NBLs look like solid masses with small areas of calcification; the presence of calcification is regarded as one of the key imaging features of NBL. More detailed imaging is necessary for predicting outcome and planning are a lot of cells packed closely together as they are in tumours.

After treatment, imaging is used to assess the degree of treatment response. Besides defining the change in size, it is important to appreciate the changes in the appearance of the tumour, diffusion characteristics and the way the tumour takes up contrast (intravenous dye) which are all regarded as additional features of response to treatment.

The tumour may become more fibrous or may calcify with treatment; sometimes it is then more difficult to see with MRI, and in particular, it may be difficult to assess the relationship of the tumour to the blood vessels. This is often better detected with CT, performed after the injection of contrast. Therefore CT can be used to show the full extent of tissue that must be removed: important for the surgeon to appreciate before surgery.

A combination of nuclear medicine and radiological examinations are crucial for diagnosis and follow-up. A type of nuclear imaging method, the I-123 MIBG scan, is used to detect hidden disease and spread of NBL to the bones. It is a method that uses a radioactive chemical that is absorbed by most NBLs.

The stage of the tumour describes its full extent throughout the body and any other features that affect the outlook for the patient. Accurate staging enables specialists to plan the treatment and give the patient's family information about what to expect in the coming months regarding treatment and side effects.

The International Neuroblastoma Staging System (INSS) is a staging system based on the imaging appearances after surgery, and is dependent on the expertise of the surgeon. The International Neuroblastoma Risk Group Staging System (INRGSS) is a

pre-surgical comprehensive staging system, which is based on the imaging appearances before surgery. Imaging is an important component of this staging system that uses well-described image-defined risk factors.

Treatment

Children with NBL are divided into three risks groups (low, intermediate and high risk) depending on age, stage of the tumour and certain biological characteristics of the tumour cells. Low risk patients have a fiveyear survival rate of >95%. The intermediate and high risk group have survival rates of 90-95% and 40-50% respectively. Management strategies include a combination of surgery, chemotherapy and radiotherapy (including MIBG therapy) with additional therapy to clear the bone marrow and more recently also immunotherapy for high-risk disease.

Future perspectives

Although no large studies have been published regarding the role of DWI in neuroblastoma, there is a potential role for this functional MRI tool in the detection and assessment of treatment response.

The use of positron emission tomography (PET), a specific sort of nuclear medicine imaging which requires a different radioactive substance and camera, is increasing in neuroblastoma, although the question remains when and in which patients it is most useful. New variations of PET imaging are currently being investigated, but all involve radiation, which can be harmful. It is important to balance the potential benefits of the additional information against that risk.

NEPHROBLASTOMA/WILMS TUMOUR

Background

Nephroblastoma (often called Wilms tumour) is a malignant tumour originating in the cells of the kidney. It is the most common type of kidney cancer that occurs during childhood. This tumour accounts for 6% of childhood cancers per year. Most children are younger than five at the time of diagnosis. Several congenital abnormalities and syndromes predispose to Wilms tumour (e.g. Beckwith-Wiedeman syndrome, and Drash syndrome). The treatment of Wilms tumour has been a success story; the cure rate is currently around 80-90%. The credit goes to the large international collaborative groups: the Société Internationale d'Oncologie Pédiatrique (SIOP) in Europe and the Children's Oncology Group in North America. Both groups have improved care of these children, but with very different approaches: The SIOP trials and studies largely focus on the issue of pre-operative therapy, whereas the COG trials and studies focused on primary surgery.

Clinical features

Wilms tumours can grow quite large before any symptoms develop. When they do appear, symptoms may be similar to those of other diseases. Most often children present with abdominal swelling, often towards one side. Sometimes there is abdominal pain or blood in the urine (haematuria). In 10% of cases Wilms tumour is discovered after coincidental trauma. High blood pressure can occur in 25% of cases and is attrib-

FIGURE 1



Four-year-old girl diagnosed with stage IV neuroblastoma with the primary tumour arising from the left adrenal gland. Coronal acquired 3D T2 image (A) with axial reconstruction (B) demonstrates the primary tumour (long arrow) with compression of the left kidney, vascular encasement (arrowheads) and lifting of the aorta (short arrow) that is considered one of the key imaging features of neuroblastoma.

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uted to an increase in hormone production by the tumour.

Imaging

Ultrasound is most often the first investigation. Complementary cross-sectional imaging (CT or MRI) of the abdomen may be required to further delineate the tumour and its surroundings. Chest x-ray (Europe) or Chest CT (North-America) is performed to assess whether the tumour has spread to the lungs.

MRI is preferred to CT since it does not use radiation and provides excellent images of the soft tissues. Wilms tumour manifests as a solid mass, usually within the kidney. The remaining part of the kidney is often stretched around the periphery of the tumour, giving the 'claw sign' (Figures 2, 3). The tumour has a mixed appearance with fluid-containing cysts, areas of bleeding and parts where some of the tumour cells have died. The tumour typically spreads by direct extension and displaces adjacent structures but does not encase or elevate large blood vessels as a neuroblastoma would (Figure 1). The tumour spreads into the veins, draining the kidney in around 6% of cases.

Ultrasound can assess the movement of the mass related to the surroundings and can be used to assess whether the internal structures next to the tumour have been invaded by it or are stuck to it.

Whenever a mass is seen in one kidney, the kidney on the other side should be carefully examined for bilateral disease that occurs in 5% of children with Wilms tumour. Spread to other areas of the body (metastasis) occurs in around 5% of children. Commonly these tumours metastasise to the lungs followed by lymph nodes or liver. In rare cases, it metastasises to the bone marrow, bone or brain.

Management

Treatment involves a combination of surgery and chemotherapy. In North America the tumour is almost always removed as soon as it is found. In Europe, the patient usually receives several weeks of chemotherapy first. However, both approaches produce equally high rates of treatment success.

For advanced disease, local radiation therapy of the tumour bed is advocated in some cases and radiotherapy to the whole abdomen can be used when there is gross tumour spillage at surgery, to decrease the risk of the tumour regrowing where the site has been contaminated.

Future perspectives

Due to the highly successful treatment regimes, the overall outcome is excellent. Future efforts will focus on reducing therapy-associated complications and minimising radiation exposure without compromising cure rates. For example, current research focuses on the issue of detecting lung metastases with chest x-rays rather than chest CT.

FIGURE 2



A four-year-old girl presented with an abdominal mass caused by a Wilms tumour arising from the right kidney. This ultrasound image demonstrates stretching of renal tissue around the tumour (also known as the 'claw sign'; arrows).

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FIGURE 3



A three-year-old girl with Wilms tumour arising from the right kidney. Coronal 3D T2 weighted image illustrates a large lobulated, heterogeneous intrarenal mass (short arrows point to the right kidney stretching around the tumour; 'claw sign') with lung metastasis (long arrows).

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BY RUTGER A.J. NIEVELSTEIN AND SUE C. KASTE

ymphoma is the third most common form of childhood cancer involving the lymphoid (or lymphoreticular) tissues of the body¹. Two major subgroups of lymphoma can be distinguished: Hodgkin Lymphoma (HL) and non-Hodgkin Lymphoma (NHL). NHL is most frequent in children younger than 15 years, whereas HL is predominant in children up to 18 years of age. In general, HL includes five disease subtypes, of which classical HL is by far the most common in children. It is one of the most curable forms of childhood cancer with an estimated five-year survival rate over 98%. On the contrary, childhood NHL involves a very varied group of mainly high-grade lymphoid cancer subtypes with variable disease courses and cure rates ranging from 65% to over 90%. A major challenge in treating children with lymphoma is to optimise up-front treatment to prevent disease relapse, while minimising late therapy-related side effects such as secondary cancers and adverse cardiac effects that may occur in adulthood. Adequate assessment of disease extent at diagnosis (staging) and response to therapy (during and at end of treatment) are essential to achieve these goals.

Children with malignant lymphoma often present with painless masses in neck, armpits or groin. If large and present in the chest or neck, they may cause shortness of breath and coughing, especially during exercise. Once lymphoma has been diagnosed (this usually involves removing one of the masses and examining it under the microscope), the extent of the disease must be determined (staging) for appropriate treatment planning and estimation of prognosis. Imaging plays a crucial role in staging and follow-up of lymphoma, by providing a non-invasive means to evaluate disease throughout the body^{1, 2}.

IMAGING TECHNIQUES

Imaging techniques used in lymphoma can be subdivided into structural and functional imaging techniques. Structural methods assess morphologic features of normal tissues and organs of the body and of malignant lesions within these structures. Functional imaging techniques provide information about tumour physiology. Typical examples of *structural* imaging



Schematic illustration of the Ann Arbor classification. Stage I disease indicates involvement of one lymph node or a group of adjacent lymph nodes; stage II, two or more nodal groups on the same side of the diaphragm; stage III, lymph nodes on both sides of the diaphragm or lymph nodes above the diaphragm with spleen involvement; and stage IV, additional noncontiguous extranodal involvement.



techniques are computed tomography (CT), magnetic resonance imaging (MRI), and ultrasound (US). Until recently, CT has been the imaging technique of choice for staging and follow-up of lymphoma. With modern (multi-slice) CT scanners, a whole-body CT scan with administration of intravenous contrast medium (contrast-enhanced CT, CE-CT) can be acquired within a few seconds, allowing for the detection of lymph nodes and other lesions even smaller than 5mm, throughout the body. A major disadvantage of CT is the use of ionising radiation, which may be associated with the induction of second cancers later in life. This small but not negligible health risk is of particular concern in children as their tissues are more radiosensitive than adults and they have more years ahead in which cancerous changes might occur. That is why alternative imaging techniques that do not use ionising radiation, such as US and MRI, are now being used more frequently.

US is a radiation-free and patient-friendly imaging technique that can provide real-time detailed images of most body parts. It can sometimes be used to image children with lymphoma, but is less useful for the evaluation of deeper-lying tissues, and for tissues located behind bones and air-containing tissues. Therefore, US is of limited use in the chest, deep retroperitoneum, and in the evaluation of obese patients. As a result, the main role of US in lymphoma is to help ascertain the nature of palpable masses and to guide biopsy procedures.

MRI uses no radiation to achieve detailed images with a high spatial resolution and excellent soft tissue contrast. This makes MRI an ideal tool for the detection of pathology, especially in parenchymal and bone marrow locations. Recent technological advances have resulted in fast, diagnostic sequences for whole-body MR imaging (WB-MRI). As a result, MRI has become a clinically feasible imaging method for staging and follow-up of malignancies, including lymphoma.

An example of a *functional* imaging technique that plays a central role in the evaluation of lymphoma is positron emission tomography (PET) using the radiotracer [¹⁸F]-2-fluoro-2-deoxy-D-glucose (FDG). Structural and functional imaging techniques are complementary to each other; by combining both techniques, anatomic localisation, lesion characterisation, segmentation, and quantification of areas with abnormal FDG uptake can be defined as in FDG-PET/CT. On the other hand, structural imaging techniques are the method of choice for identifying relevant space-occupying consequences of lymphomatous masses such as airway and vascular compression/obstruction, spinal cord compression and bone lesions at risk of fracture.

STAGING IN LYMPHOMA

Lymphomas are usually staged according to the Ann Arbor staging system or recently introduced Lugano modification (Figure 1)³. The Ann Arbor staging system divides patients into four stages based on localised disease, multiple sides of disease on one side or both sides of the diaphragm, and disseminated extranodal disease. Paediatric NHL are usually staged using the St. Jude staging system which shows great similarity to the Ann Arbor staging system but better takes into account the more frequent presence of extranodal disease (such as the gastrointestinal tract, solid abdominal viscera and bone marrow). CE-CT is still the most



PET/CT in a boy, 15 years of age, with a Hodgkin Lymphoma (nodular-sclerosing type). A: Whole body Maximum Intensity Projection (MIP) overview of the PET acquisition illustrating the FDG radiotracer uptake in the involved lymph nodes in thorax and abdomen (black arrows). B: Axial fused PET/CT image illustrating the better anatomical delineation of the FDG radiotracer uptake in the enlarged abdominal lymph nodes (white arrows). C: Corresponding diagnostic (contrast-enhanced) CT image illustrating the enlarged lymph nodes in the abdomen (white arrows). D: Post-treatment whole-body MIP overview of the PET acquisition showing a small focus of residual FDG radiotracer uptake in the abdomen (black arrow). In the thorax, a small residual amount of FDG tracer is visible in the reservoir of the central venous line used for the administration of the radiotracer (arrowhead). E: Axial fused PET/CT image illustrating the better anatomical delineation of the FDG radiotracer uptake in a residual enlarged lymph node (white arrow). F: Corresponding diagnostic (contrast-enhanced) CT image illustrating residual enlarged lymph nodes in the abdomen, in between the inferior vena cava and aorta (white arrow). Because of this small focus of residual FDG radiotracer uptake in the abdomen (and mediastinum, not shown) after chemotherapy, the patient will receive additional radiation therapy to those sites of persistently active disease.

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commonly used structural imaging technique for staging of lymphoma. However, functional imaging using FDG-PET or FDG-PET/CT for staging (baseline PET) is becoming the norm. Until recently, FDG-PET or FDG-PET/CT had been reserved for therapy evaluation of lymphoma, but several recent studies have shown that baseline PET facilitates and influences the interpretation of a post-treatment PET in a considerable number of patients. Thus, current international guidelines recommend the use of (integrated) FDG-PET/CT in staging of lymphoma. However, because of the health risks related to the use of ionising radiation, there is increasing interest in developing WB-MRI techniques for staging and follow-up. Several recent studies have shown that WB-MRI is feasible, even in children, and the agreement between WB-MRI and FDG-PET/CT reference standard in staging lymphoma is good⁴. Therefore, WB-MRI may become an attractive alternative to CE-CT and FDG-PET/CT in this radiosensitive population, at least for staging purposes.

THERAPY RESPONSE EVALUATION IN LYMPHOMA

The evaluation of response to therapy during and at the end of treatment is of great importance for determining the effectiveness of treatment, the need for additional treatment, and the prediction of clinical outcome. Identification of non-responders at an early stage during treatment allows the treatment strategy to be adjusted and thereby may improve prognosis and minimise late therapy-related side effects. Although the value of altering therapy based on early or mid-treatment imaging in children remains to be established, recent studies in children already suggest that interim FDG-PET or FDG-PET/CT during therapy is an excellent prognostic indicator for predicting clinical outcome.

Imaging based evaluation of response to therapy is performed according to the revised Cheson criteria (or Lugano classification) for response assessment in lymphoma³. These criteria are based on an evaluation of both CE-CT and FDG-PET/CT imaging, whereby CE-CT is reserved for evaluation of variable and non-FDG-avid types of lymphoma, distinguishing bowel from lymphomatous masses, and evaluation of compression/thrombosis of central vessels and airways. According to the Cheson criteria (Lugano classification), complete remission indicates the disappearance of all evidence of disease, partial remission indicates the regression of measurable disease and no new sites, stable disease indicates failure to attain complete remission or partial remission, and progressive disease indicates the appearance of new lesions or increase by ≥50% of previously involved sites. The role of WB-MRI in therapy response evaluation of children with lymphoma is under investigation.

CURRENT RESEARCH AND FUTURE PERSPECTIVES

Where imaging was initially regarded and used mainly as a tool to visualise anatomy and structural changes in the body (i.e. to detect tumours and to assess the anatomic extent of tumour spread before and after initiation of therapy), it is increasingly used to gain information on the biological behaviour of tumours



(black arrows).

CHAPTER 6: PAEDIATRIC CANCER IMAGING





WB-MRI in a boy, 15 years of age, with a Hodgkin Lymphoma, stage II. A and B: Coronal T1-weighted and STIR images illustrating the massive lymph node enlargement in the left and right cervical regions and mediastinum (white arrows). C: On the DWI the pathological lymph node masses are highlighted due to the pathological diffusion restriction in the lymph nodes and the background suppression of normal anatomical structures

(i.e. functional imaging and use of imaging biomarkers). Therefore, current research focuses on developing objective imaging-based measures (imaging biomarkers) for better therapy planning and response assessment, as well as better prediction of clinical outcome. Most current studies are evaluating the role of FDG-PET in these perspectives, and include the introduction of new (more tumour-specific) radiotracers and quantification of biological behaviour. However, there is an increasing interest in investigating the potential role of MRI, particularly advanced MR imaging techniques like diffusion weighted imaging (DWI) and magnetic resonance spectroscopy (MRS). DWI allows for non-invasive visualisation and quantification of the random microscopic movement of water molecules within biological tissues². One of the main advantages of DWI over conventional MRI sequences is its ability to highlight lesions while sup-

pressing signal from many unwanted background tissues such as fat, flowing blood, cerebrospinal fluid, and gastrointestinal contents. Furthermore, the ability of DWI to quantify diffusion in biologic tissues by means of apparent diffusion coefficient (ADC) measurements may aid in the characterisation and treatment response assessment of lymphomatous lesions. MRS allows for separation of the MRI signal from a given tissue into its different chemical components, which may improve lesion characterisation and prediction of clinical outcome. In this scope, it is interesting to mention the recent development of integrated PET/MRI systems, which combines the strengths of both imaging techniques while considerably reducing the radiation dose to the child. At present, only a limited number of (experimental) PET/MRI systems have been developed and are being tested worldwide.

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Where imaging was initially regarded and used mainly as a tool to visualise anatomy and structural changes in the body [...], it is increasingly used to gain information on the biological behaviour of tumours [...]. Therefore, current research focuses on developing objective imaging-based measures (imaging biomarkers) for better therapy planning and response assessment, as well as better prediction of clinical outcome.

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MUSCULO-SKELETAL IMAGING



BONE TUMOURS

BY CLAUDIO GRANATA

Bone tumours in children may be either benign or malignant. A malignant tumour is a growing mass of cells that has the ability to invade the structures around it and may spread (metastasise) to affect distant parts of the body. A benign tumour grows locally, where it may cause problems, but it does not invade the surrounding tissues and cannot spread.

The majority of primary bone tumours are benign; they may not cause any symptoms and can remain undetected or be noticed by chance when radiographic examinations are performed for other reasons. Common benign bone lesions include non-ossifying fibroma, bone cysts, osteochondroma, Langerhans cell histiocytosis, osteoid osteoma, and aneurysmal bone cysts. Occasionally, some of these conditions may weaken bone structure, causing fractures which appear to occur spontaneously or after only minor trauma, such as at a school sports day.

Malignant bone tumours – mainly osteosarcomas and Ewing's sarcoma – are fairly rare in children, accounting for about 5% of all childhood malignancies. Pain is usually the main symptom, which is often overlooked until a fracture or local swelling occurs. Other non-cancerous conditions, such as bone infections, stress fractures (fractures caused by overuse), and some conditions that affect joints, may simulate a primary malignant bone tumour.

Despite the great advances in diagnostic imaging with the introduction of computed tomography (CT) and magnetic resonance imaging (MRI), conventional radiography (x-ray) remains the mainstay for diagnosis and differential diagnosis of bone tumours. A wealth of information can be acquired with radiography, which is the first imaging modality that should be performed. X-rays can identify the site of the bone abnormality, features of bone destruction by the tumour, and the extent of involvement of adjacent soft tissues. They can also show how the fibrous covering around the bone (the periosteum) is responding to the tumour, which is known as the periosteal reaction.

Benign lesions are usually characterised by areas of bone destruction with well-defined borders; the periosteal reaction – if any – is solid and uninterrupted, and adjacent soft tissues are not involved. On the other hand, bone destruction with ill-defined borders, interrupted periosteal reaction, and adjacent soft tisD





A peripheral lesion in the distal tibia. Its characteristics (rounded, well defined, elliptical shape, lucent centre) and absence of worrying features suggest that it is a benign lesion, probably a nonossifying fibroma.

sue involvement is observed more often in malignant tumours. Nevertheless, in some instances it can be very difficult to differentiate a benign from a malignant condition with radiography alone.

MRI is considered the second examination to be performed in cases of suspected malignant bone tumours. MRI is helpful for defining the local extent of the tumour, as it provides excellent tissue contrast and detailed visualisation of bone marrow and soft tissue involvement. When a bone tumour contains specific tissue components, such as fat, fluid, blood and blood vessels, or cartilage, MRI imaging may show characteristics of a particular type of tumour, leading to a specific diagnosis. Further information about the tumour can be obtained by the injection of contrast medium.

CT is a complementary technique, which can be useful in depicting bone involvement and periosteal reaction, especially in structures with complex anatomy such as head, face, pelvis, or scapula, where images can be reconstructed in multiple planes and even in 3D. CT can also be useful in assessing tumours with a cartilaginous content, and in guiding interventional bone biopsies. Some tumours (osteoid osteoma) have highly characteristic features on CT which enable the diagnosis to be made with certainty.

CT is also better than a chest x-ray for the detection of lung metastases in malignant bone tumours, although the benefits of its use (the expected amount of useful information) have to be balanced against the relatively high dose of radiation.

Bone scintigraphy (also known as a bone scan) detects bone areas where there is increased activity of the cells which produce new bone (osteoblasts) associated with change and repair. This technique is very sensitive for spotting bone tumours, but it is usually not possible to use it to differentiate benign from malignant lesions and tumours from tumourlike lesions. Bone scintigraphy is especially useful for the detection of multiple skeletal lesions, which can be observed in some childhood conditions such as Langerhans cell histiocytosis or fibrous dysplasia.

Despite the wealth of information which can be collected with the imaging modalities mentioned above, the precise diagnosis of a bone lesion may remain difficult to obtain because imaging findings are often not specific enough to make a particular diagnosis, and many different bone lesions, including tumours, infection and some metabolic conditions, can have similar or overlapping features. In these cases it is usually necessary to obtain a small sample of the abnormal area (a biopsy) and examine it under a microscope. Imaging may be used to guide this procedure and ensure the correct part of the bone is biopsied.

The integration of biopsy findings, clinical data, and imaging information is of utmost importance in reaching the final diagnosis. Close cooperation between orthopaedic surgeon, paediatric radiologist, and pathologist is essential.



A radiograph of a peripheral lesion in the distal femur (A). Its x-ray characteristics (bone destruction, illdefined margins, periosteal reaction) as well as surrounding oedema (B; T2 weighted MRI) and patchy contrast enhancement (C; contrast enhanced T1 weighted MRI) suggest a malignant lesion: osteosarcoma.

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IMAGING CHILDHOOD ARTHRITIS

BY KAREN ROSENDAHL

rthritis is an inflammation of the joints that is characterised by swelling, heat, and pain. It can be short-term, lasting for just a few weeks or months, or it can be chronic with ongoing activity into adulthood. In some cases, it can last a lifetime. However, with modern treatment and support, disease activity is manageable and children can lead full and active lives. The commonest form of childhood arthritis is juvenile idiopathic arthritis (JIA), affecting about 1 to 2 per 1,000 children under the age of 16 years, or more than 50,000 children in the United States.

WHAT CAUSES JIA?

It's not known exactly what causes JIA, but research suggests that it is an autoimmune disease, and that environmental as well as genetic factors may play a role. In autoimmune diseases, instead of the immune system protecting the body, it releases chemicals that can damage healthy tissues and cause inflammation and pain. To effectively manage and minimise the effects of arthritis, an early and accurate diagnosis is essential. This is where modern imaging can make a difference.

TYPES OF JIA

JIA usually first starts in children between six months and 16 years of age. Although there are seven major types or categories of JIA, this classification may not be helpful at the beginning of the disease, as it does not help to predict which children are most likely to develop joint damage, and who therefore require a more active treatment at an early stage. Again, modern imaging can make a difference.

The seven subtypes are as follows 1-3:

1. Oligoarthritis

This is the most common type, affecting 27-56% of children with JIA. Beginning in early childhood, typically four or fewer joints are involved, the knee and wrist joints most commonly. The two types of oligoar-

2. Polyarticular arthritis, rheumatoid factor negative

Affects 18-28% of children with JIA; symptoms include swelling or pain in five or more joints. Usually presents during early childhood, with a second peak at 6-12 years. The small joints of the hands are affected as well as the weight-bearing joints like the knees, hips, ankles, feet, and neck.

3. Polyarticular arthritis, rheumatoid factor positive

Affects 2-7% of all children with JIA; presents in late childhood or adolescence. The disease resembles adult rheumatoid arthritis with involvement of small joints.

4. Psoriatic arthritis

Affects 2-11% of children with JIA; children with this subtype have psoriasis rash, or a close relative with psoriasis. Controversial definition, resembles oligoarthritis but more often including affected fingernails or toenails, and involvement of small and large joints.



thritis, persistent and extended, are determined by how many joints are involved over time.

5. Enthesitis-related arthritis

Affects 3-11%, presents in late childhood or adolescence and most commonly affects the lower extremities and the spine. Children might also have inflammation where tendons join bones. Enthesitis-related arthritis includes a special group called juvenile ankylosing spondylitis (where joints of the low back are inflamed) and arthritis associated with inflammatory bowel disease (Crohn's disease and ulcerative colitis).

6. Systemic JIA

Affects the whole body (4-17% of all children with JIA). Symptoms include high fevers that often increase in the evenings and then may suddenly drop to normal. During the onset of fever, the child may feel very ill, appear pale, or develop a rash. The spleen and lymph nodes might become enlarged. Eventually many of the body's joints are affected by swelling, pain, and stiffness.

7. Undifferentiated arthritis

Arthritis that doesn't fit into any of the above categories or fits into more than one of the categories.



DIAGNOSIS AND TREATMENT

To diagnose JIA, the doctor will take a detailed medical history, perform a thorough physical examination and order blood tests and imaging examinations to rule out other conditions or infections, particularly when only a single joint is affected. Treatment includes a combination of medication, physical therapy, and exercise. Historically, nonsteroidal anti-inflammatory drugs (NSAIDs) were used as the first line of treatment for JIA, but evidence now shows that earlier and more aggressive treatment with disease-modifying anti-rheumatic drugs (DMARDs, including new biologic drugs) can improve disease control and long-term outcomes, provided that the conditions with high risk of severe outcome can be predicted early and appropriate treatment instigated. However, response to the novel, biologic agents varies across patients and needs to be monitored closely and adjusted as required, to optimise benefits and to reduce the unintended risks.

THE ROLE OF IMAGING IN JIA

Imaging plays an important role in the diagnosis of children with JIA, particularly of joints that are frequently affected but may have no or very few symptoms, such as the joint of the jaw (the temporomandibular joint, TMJ). Insufficient treatment of the TMJs may lead to abnormal growth around puberty and restricted growth of the jaw. Imaging is used to determine the degree and extension of joint inflammation, to evaluate treatment response and to determine whether or not permanent joint damage has occurred.

TYPES OF IMAGING

Traditionally, joint damage evaluation has been performed with plain radiography (x-rays). The method can show bone destruction (or erosion), which occurs relatively late in the disease course, cartilage loss (indirectly, through joint space narrowing), and joint misalignment, but it cannot visualise the active, inflammatory change directly (Figure 1). Plain radiographs have particularly low sensitivity for disease in early stages. Nevertheless, they are often used as a baseline examination, and to rule out other bone disease. For the temporomandibular joint, cone beam computed tomography (CBCT) is currently accepted as the best type of imaging for visualisation of bony structures (Figure 2).

In contrast, ultrasound can visualise inflammatory change such as increased joint fluid (joint effusion) and a thickened synovial membrane (joint lining) (Figure 3). Ultrasound may also be used to guide joint aspiration for diagnostic purposes, and for therapy, e.g. tendon sheath or joint injection. The method is, however, not very accurate when it comes to diagnosing permanent damage, or bone destruction.

Magnetic resonance imaging (MRI) on the other hand, is able to show active inflammation, e.g. a thickened, inflamed synovial membrane and bone marrow oedema (increased water content within the bone marrow), as well as damage to cartilage and bone, and is believed to detect erosive changes with greater sensitivity than radiography, particularly in early disease (Figure 4). Findings suggestive of early disease are of particular interest, and it has been proposed that bone marrow oedema represents a precursor of erosive change.





Ultrasound image of the left wrist, sagittal view, in a nine-year-old girl with JIA, showing an effusion and synovial thickening in the radiocarpal joint (arrows) and in the midcarpal joint (open arrows).

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X-ray of the left hand in a four-year-old girl with JIA, showing swelling of the 4th finger (arrows) and an inflammatory reaction to the bone (arrowheads).

FIGURE 2



Cone beam computed tomography (CBCT) of the temporomandibular joints in a twelve-year-old girl with JIA. On the left side there is a flattened and irregular condyle (A, arrow), while the right joint is normal (B).

The value of MRI, as an advanced method for evaluating disease activity and disease damage in adults with rheumatoid arthritis, is under active investigation by a research consortium called Outcome Measures in Rheumatology Clinical Trials (OMERACT). However, the results that will be drawn from OMERACT studies are not directly applicable to children, because adult rheumatoid arthritis is different from JIA and because the growing skeleton needs a different approach⁴. Indeed, in children, the bony surfaces can be surprisingly irregular, thus mimicking erosive JIA-changes (Figure 5). Moreover, some of the MR-appearances of immature bone marrow may be mistaken for bone marrow oedema and misinterpreted as active inflammation.

Currently there are no unifying international recommendations for imaging in JIA and both the choice of imaging and who and when to refer for radiological investigations vary between centres^{2, 4}. One of the major challenges when interpreting imaging studies is to distinguish between normal findings and pathology in the early stages of the disease, particularly on MRI (Figure 5)⁴. Moreover, the lack of standardised scoring systems for all the radiological imaging methods makes the objective evaluation of the degree of inflammation and destruction challenging.

In sharp contrast to the level of investment and consequent advances in research addressing the adult disease (rheumatoid arthritis), little is known about advanced imaging markers for active arthritis and permanent damage in children with JIA. This has fuelled a large, European multicentre study on imaging in JIA, which aims to develop precise and valid, child-specific imaging markers and scoring systems to allow for evidence-based clinical practice as well as robust drug trials. Furthermore, novel guidelines for referral and standards of practice for imaging in JIA will reduce the risk of inappropriate and unnecessary referrals and examinations, with the potential to reduce radiation exposure.

The research group includes highly experienced and well-known paediatric radiologists, rheumatologists, dentists, oral surgeons, and radiographers, as well as medical physicists and mathematicians, representing several of the active research groups within this particular field, and possessing a unique range of expertise⁵. Our ultimate goal is to help doctors diagnose JIA at an early stage, so that the appropriate treatment can be instigated with the potential to reduce the risk of permanent joint damage, disability and reduced quality of life.



(arrows). h = femoral head.

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MRI of the left hip joint in a ten-year-old boy with JIA, A) before intravenous contrast administration and B) after contrast, showing an inflamed joint



MRI of the left wrist in a ten-year-old boy with JIA (A), showing a bony defect in one of the carpal bones (arrow), as compared to a similar defect in a healthy ten-year-old (B), most likely representing normal variation.

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IMAGING ABUSE IN INFANTS

BY RICK R. VAN RIJN AND THOMAS L. SLOVIS

ust a Friday morning at an Emergency Department, somewhere in the world, an ambulance ✓ arrives with a four-week-old boy. He has a painful swollen right leg, noticed during diaper changing. After a physical examination by the emergency department doctor a radiograph is obtained which shows a fracture of the femur (broken upper leg bone). According to the stepfather he was alone with the baby, at home playing a computer game. When it was time to feed the baby and change his diaper he noted the swollen and painful leg. The stepfather has no explanation for the fracture. In this setting, trauma to a non-mobile baby without an independent witness, the question arises: what happened; was it an accident, delayed detection of a birth trauma or did someone hurt this infant?

Most of us see children as one of the most, if not the most, precious gifts of life and, as a result, we cannot imagine that someone would in any way abuse a child. Unfortunately the sad fact is that child abuse is real for too many children. In 1999 the World Health Organization stated 'Child abuse or maltreatment constitutes all forms of physical and emotional ill-treatment, sexual abuse, neglect or negligent treatment or commercial or other exploitation, resulting in actual or potential harm to the child's health, survival, development or dignity in the context of a relationship of responsibility, trust or power'. The incidence of child abuse varies from country to country and even within countries, but according to data from the World Health Organization (WHO), globally a guarter of all adults state that they have been physically abused as a child. Research by Vincent Felitti has shown that, not only at the time of abuse but even as an adult, the child abuse carries long-term serious medical and psychological consequences. Besides the medical and social implications, child abuse also leads to a high cost to society. In 2012, the Centers for Disease Control and Prevention calculated that, based on 1,740 fatal and 579.000 non-fatal confirmed cases of child abuse, the costs for both direct medical care as well as long-term consequences such as adult medical costs, loss of productivity, and special education costs, were a staggering 124 billion US Dollars each year.

What role do paediatric radiologists play in managing child abuse? They are part of the medical team of specialists who evaluate the radiographs and other images that raise the suspicion of abuse. They may be a skeletal survey).

handling of a child.

In abused children, skeletal injuries are seldom life threatening but they often represent the most convincing signs of abuse on x-rays. While certain fractures, in otherwise healthy children, can be perceived as almost characteristic of child abuse it should be stressed that any fracture can be caused by abuse. The fracture is proof that the child has been injured. In the end, the medical team suggests that there may be abuse but it is up to the courts to decide who caused the injury, and what the mechanism of the injury was. The role of the paediatric radiologist, as part of a medical team of specialists, is to evaluate the imaging examinations in the light of the explanation provided.

Paediatric Imaging

the first physician to suggest abuse based on the kind of injury or discrepancy between the injury and history. The paediatric radiologist may also suggest further studies (such as a detailed series of radiographs;

The incidence of physical abuse is highest in infants, and fractures are the most frequent injury after bruises. The most severe injuries are to the infant brain (abusive head trauma), and fractures and abusive head trauma frequently occur together. The importance of radiology in the diagnosis of physical child abuse is underlined by the fact that when, in 1946, the paediatric radiologist John Caffey, MD (1895-1978) published his innovative paper *Multiple fractures in the* long bones of infants suffering from chronic subdural hematoma (blood between the brain and skull), he was one of the first to cautiously include maltreatment against children as a possible cause of fractures. In this initial paper, no other signs of bone disease, such a scurvy, rickets, or lead poisoning, were present. These are diseases that, although rare now, were common in previous generations and may have caused weak bones, which could lead to fractures during normal

The first task of the radiologist is to evaluate and describe the appearance and location of the fracture. Certain fractures raise more suspicion of child abuse, the most significant being fractures at the back of the ribs (posterior rib fractures) and metaphyseal corner fractures (fractures at the end of long bones) in children under the age of two years. Posterior rib fractures are thought to be the result of compressing the chest, leading to fractures of the ribs by the gripping forces of the fingers. These fractures become easier to identify when they heal and an extra amount of bone (known in radiology as callus) becomes visible. They are sometimes diagnosed in children who undergo a chest x-ray for other reasons, such as to rule out pneumonia. In these cases the paediatric radiologist is usually the first doctor to suggest the diagnosis of child abuse. These fractures can of course also be seen on computed tomography (CT), but CT involves a higher dose of radiation than an ordinary x-ray, so this is not a standard approach. Another fracture which should raise the diagnosis of child abuse is the metaphyseal corner fracture, also known by the name given by P.K. Kleinman: classic metaphyseal lesion. These fractures are located at the edges of long bones and are the result of twisting or pulling on the arm or leg.

If a child has multiple fractures occurring at the same time or if there is evidence of several episodes of trauma, this also raises concerns of child abuse. One of the tasks of the paediatric radiologist is therefore also to try to 'date' fractures, i.e. to estimate the time interval between the moment the fracture occurred and the moment the radiograph that was taken. Although exact dating is not possible, radiologist are able to tell the difference between fresh fractures, fractures which show the first sign of healing, and fractures that are already healing. This information can be essential in helping to make the correct diagnosis.







Fracture of the left upper leg of a five-month-old boy. The infant suffered from multiple fractures without a proper explanation. After confronting the parents with the presence of these fractures, the father admitted to abusing the infant.



Radiograph of the left knee of a two-month-old infant. According to the parents, he became unconscious during a bottle feed and after a few minutes he regained consciousness but was less alert then before the incident. After presentation at the emergency department a skeletal survey was performed which showed the metaphyseal corner fracture shown in this image. A fracture of the right upper leg was also found.

Coronal MRI of a five-month-old infant. According to her stepfather she suddenly became unconscious while in his care. Magnetic resonance imaging shows bilateral subdural haematomas (blood between the brain and the skull). After interrogation by the police the stepfather admitted abuse.

References



FIGURE

Paediatric Imaging

However, not only the fracture but also the general appearance of the affected bone should be assessed. It is important to be aware of potential underlying diseases, e.g. metabolic disorders (diseases in which bone formation is hindered) or inherited bone disease such as Osteogenesis Imperfecta (a collagen disorder with an increased risk of fractures, sometimes known as 'brittle bone disease'). When examining the x-rays the radiologist should be aware of findings which could suggest an underlying disease and, if found, these should be discussed with the physician looking after the child, so that follow-up blood tests or other examinations can be performed.

Besides fractures, trauma to the abdomen or brain (known as abusive head trauma) can also result from child abuse. Abdominal trauma in children is relatively uncommon but, due to its delayed presentation, it carries a high mortality rate. The role of radiology in these cases is to ascertain which organs are affected. In most cases, CT will be used to identify and assess abdominal trauma. With respect to abusive head trauma, the role of the paediatric radiologist is to

interpret and report the findings from CT and magnetic resonance imaging (MRI). The presence of subdural haematomas, i.e. blood between the brain and the skull, without an adequate explanation is a strong indicator of abusive head trauma. However, other injuries to the brain can also be the result of abusive head trauma. Besides identifying brain lesions, the paediatric radiologist also uses MRI to assess the neurological outcome of the child, i.e. to what extent will the child suffer permanent brain damage.

In summary, paediatric radiologists play a crucial role in the detection of physical child abuse. The input they provide in comparing radiological findings with the information provided by the parent or caregiver is paramount. Can the trauma described and the mechanics related to it result in the injuries seen on imaging? The role of the paediatric radiologist is that of the indispensable imaging expert using the full range of radiological imaging methods, such as radiographs, CT, MRI and ultrasound, both with respect to interpreting as well as in advising additional imaging, in a multi-disciplinary child advocacy team.

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PERINATAL IMAGING

TAL IMAGING



FOETAL IMAGING

BY DOROTHY I. BULAS AND FRED E AVNI

INTRODUCTION

Ultrasound scanning was first used 40 years ago to produce pictures of unborn babies in the womb and is still used all over the world. It is safe for the baby, relatively cheap and widely available. In many places paediatric radiologists are involved in performing and interpreting these scans. This can be very helpful if abnormalities are discovered because paediatric radiologists have experience of looking after babies and older children, and have an excellent understanding of what implications there may be for the child and their families in the long term. The radiologist is responsible for informing other specialist doctors about the scans and can play a vital role in coordinating the care of the mother and baby.

An ultrasound scan performed in pregnancy can show the number of babies, what position they are lying in and whether everything looks normal (Figure 1). Occasionally operations can be performed on unborn babies in the womb, and ultrasound is used to guide these highly specialised procedures. The quality of the ultrasound pictures is however, dependent on the skill and experience of the person performing the scan and the rate of detection of abnormalities varies from 13-82%. Some conditions can be extremely difficult to detect with ultrasound even by very experienced operators. Other factors affecting the quality of the scan are the position of the baby and the amount of sedation for foetal MRI is rare.

fluid around them. The size and weight of the mother also have an influence.

In recent years, magnetic resonance imaging (MRI) has advanced a great deal, and scans which used to take a long time can now be performed in seconds, meaning that it can also be used to examine the unborn child (Figures 2, 3, 4). Paediatric radiologists have played an important role in adapting MRI techniques so that it is suitable for use in pregnancy.

HISTORY OF FOETAL MRI

In the past, MRI was not used to image the foetus; the scans took so long to perform that the babies usually moved around and the images then became too blurred to be helpful. It was first used in the 1980s, originally just to look at the size of the foetus and to assess the placenta. These scans were performed in several short bursts of 2-3 minutes each.

Initially, in order to stop the foetus moving around so much, paralysing drugs were injected into the baby inside the womb via the umbilical cord, but this was rapidly stopped and instead sedative drugs were given to the mother, which then affected the baby. Nowadays, scans are so much faster that the need for outlook and implications.

In the 1990s, MRI was used to assess babies in whom the ultrasound scan indicated the diaphragm had not developed properly, which was another important step forward. Not only was the technique used to diagnose the abnormality, called diaphragmatic hernia, and to show whether other body parts such as the liver had been displaced, but it was also used to look at the size of the lungs, which is important in assessing the outlook for the babies.

By 2000, further improvement in the technique allowed the study of the bowel by assessing meconium (foetal stool). MRI techniques continue to advance all the time, and new possibilities are always developing.

MRI ADVANTAGES

MRI and ultrasound differ in several ways that makes the two techniques complementary. MRI, unlike ultra-



In the late 1980s and early 1990s, advances in technology enabled MRI to be used much more frequently to assess unborn babies. It proved to be much better than ultrasound at assessing abnormalities of the brain and radiologists were able to see these more clearly and discuss them with the appropriate specialist doctors who could then advise the family about the



Three-dimensional ultrasound of a foetal face at 24 weeks gestation.

sound, is not limited by the position of the baby overlying maternal or foetal bone, obesity, or reduced amniotic fluid.

Advanced MR imaging has been particularly useful in imaging abnormalities of the foetal brain, which can be limited with ultrasound as it cannot pass through the bony skull.

The foetal airway is filled with fluid, allowing MRI to directly visualise the back of the throat and the windpipe – regions not well visualised by ultrasound. This is particularly important when there is a lump in the mouth or neck, compressing the airway.

MRI images are easier for the non-specialist to understand and make it easier for all the team looking after the patient to plan treatment and surgery if necessary. During counselling, families can review these images with the medical team, which is useful in understanding how recommendations have been determined. Additional benefits include the fact that foetal MRI may limit the need for postnatal imaging, which is particularly helpful when a newborn is unstable and unable to tolerate sedation.

TIMING AND INDICATIONS FOR FOETAL MRI

For the reasons stated above, MRI can be a useful addition when a targeted ultrasound scan raises questions that could benefit from further evaluation. MRI may provide additional information when an abnormality is identified by ultrasound or when findings are difficult to interpret.

The ideal timing of a foetal MRI depends on what questions need to be answered. In early pregnancy, foetal MRI is limited due to the small size of the foetus and the fact that these small foetuses move around a lot. Also, some abnormalities may have not yet developed. MRI in the middle of the pregnancy, between 18 and 22 weeks, can be useful for confirming findings identified on routine ultrasound scans or for assessing them further.

MRI in the last third of pregnancy is particularly helpful for the assessment of brain abnormalities. At this later stage the foetus has developed sufficiently for them to become much more visible, however, these later exams risk identifying anomalies too late for anything to be done. Centres may elect to perform an MR in the second trimester and then a follow-up exam in the third trimester in cases when complex delivery planning is indicated (such as large lumps in the neck).

Foetal MRI has helped the development of new surgical techniques where operations are performed on the foetus in the womb for conditions such as spina bifida and rapidly growing tumours. Accurate imaging for planning the surgery is critical in operations where both the foetus and mother are at risk.

Foetal MR images can be useful for delivery planning and postnatal surgery in cases of tumours and conjoined twins that may require surgery immediately following their delivery.

FIGURE 2



MRI performed in 1989 for a foetal mass. The mass turned out to be a Wilms tumour (image provided by Paolo Toma, MD).

CHAPTER 8: PERINATAL IMAGING





FIGURE 3



Foetal MRI image of a foetus at 22 weeks gestation.





MRI image of a foetal face and hand at 28 weeks gestation.



Foetal CT scan of a foetus in the third trimester with a skeletal dysplasia (image provided by Marie Cassart, MD).

FOETAL COMPUTED TOMOGRAPHY

formed.

Foetal CT scans are performed with such a low radiation dose that only the foetal bones (not the internal organs) are seen. Images are displayed in three dimensions, showing the foetal skeleton detail

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Although it is best to avoid imaging which uses radiation in foetuses and young children, low-dose foetal computed tomography (CT) can be used to evaluate the foetal skeleton. This is limited to special cases when severe bony abnormalities are already suspected after an ultrasound examination, yet the diagnosis is still in question and further information would help to decide whether the pregnancy should be allowed to continue. Foetal CT should only be performed in the second or third trimester, when the baby's organs are already

that currently is not achievable either by ultrasound or MRI (Figure 5). The paediatric radiologist can analyse the skull, shoulders, ribs, vertebral column and limbs, helping to identify specific skeletal abnormalities.

CONCLUSIONS

Expertise in MR, CT and ultrasound, familiarity with genetic syndromes and congenital defects, as well as an understanding of the evolution and long-term implications of abnormalities place the paediatric radiologist in a unique position of bridging the foetus with a team of advanced specialists who may provide critical care before, during and after birth. This exciting and rewarding field in paediatric radiology has provided rich opportunities for research, and the chance to support families with up-to-date technology in a compassionate manner.



THE FUTURE OF PAEDIATRIC RADIOLOGY RESEARCH

BY OWEN J. ARTHURS AND PETER J. STROUSE

esearch is key to the future of paediatric radiology. Investment in well-designed research tri-Als to address today's challenges and guestions will improve the healthcare of children tomorrow.

Clearly, resources are limited, so time and funding must be prioritised to address the most urgent and far-ranging problems. Areas of current study which warrant further work include cancer imaging in children, management of radiation exposure in paediatric imaging (both by improving dose efficiency in modalities requiring ionising radiation and also by improving alternative imaging modalities to reduce dependency on radiation-based imaging techniques), and the application of advanced new imaging methods to complex paediatric disease.

The methods which physicians currently use to manage disease will change dramatically in the next 10 or 20 years. Undoubtedly, image-guided therapy (radio-labelled antibodies, ultrasound-aided micro-bubble delivery of drugs) will revolutionise treatment of common disease processes. Other developments will change how and why imaging is performed. For example, if researchers discover a blood test which predicts survival from a certain tumour, then imaging methods currently used to predict outcome will become obsolete. Instead, imaging techniques could be used to investigate the response to treatment or to plan surgery or an interventional radiology procedure. Equally, if researchers find a cure for a certain tumour, irrespective of where it has spread to, then our current imaging methods of evaluating spread will no longer be necessary. Such major breakthroughs are rare and take time. In the meantime, our imaging methods of diagnosis need to evolve.

Current experience tells us that children with the same kind of tumour often differ in response to the same treatment. Imaging is often used to try to improve the quality of life of children by helping to more accurately predict their outcome. We are learning that tumour genetics vary between different patients and even within the substance of a single tumour in the same patient. 'Radiogenomics' is the term applied to novel imaging methods used to detect genetic variation within tumours that may predict response to therapy and guide treatment decisions. Several laboratories around the world are working on cell-based imaging markers, which may be able to identify which cells are problematic and provide a new way of targeting treatments. If we could precisely measure or predict these tumour responses using a new imaging method, which could tell which cell type was dominant within a tumour, then we might be better able to direct therapy so as to maximise the desired response but minimise its local and systemic side effects.

In recent years, it has become increasingly apparent that there are numerous genetic syndromes which predispose children to cancer. We need to learn how best to diagnose children with cancer predisposition syndromes and how to monitor them most effectively for tumours, bearing in mind that the underlying syndrome may render them more susceptible to the side effects of radiation.

It is likely that we will continue to depend on traditional x-ray and computed tomography (CT) imaging methods, which use ionising radiation. Radiation-based technologies are widely used by all hospitals around the world (only excepting some in underserved third world nations with no imaging at all).

There is considerable debate in the current literature on the true deleterious nature of ionising radiation as a cancer-causing agent. The lifetime study of cancer later in life.

Radiation biology remains an interesting field warranting further investigation. In the meantime, our attention remains focused on studying how we can make our current imaging methods more efficient. This includes reducing the radiation dose of modalities using ionising radiation (x-ray, fluoroscopy, CT, nuclear medicine) while maintaining or improving image quality and improving the quality and efficiency of modalities which do not use ionising radiation (ultrasound, magnetic resonance imaging [MRI]). In children, considerable research effort is being invested in reducing radiation exposure in CT, and to improving MRI to make it faster and more capable of diagnosing more conditions. Current attention is also focusing on possible long-term deleterious side effects of sedation or anaesthesia. Research to lessen the need for sedation and anaesthesia for MRI could prove very fruitful.

One of the most challenging and frustrating aspects of paediatric disease are rare diseases. Many paediatric conditions and certain tumours will affect only a handful of children across the world, and therefore each institution may only be able to manage and treat one or two children with each condition. Research is hampered by small numbers and limited funding.

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atom bomb survivors from the 1940s and of other radiation exposed populations (i.e. British radiation workers) suggest that low levels of radiation similar to those used in CT may slightly increase the risk of cancer later in life. However, in 2015, we are only currently beginning to see results of retrospective studies of large patient populations exposed to radiation by diagnostic imaging. We do not fully understand the effect of radiation on DNA and do not know the degree, if any, to which low levels of radiation used in diagnostic imaging increase the risk of developing

Working together across institutions, we should be able to share information and work together to determine better methods of diagnosis and better treatment protocols for rare paediatric diseases. Advances in information technology, imaging/clinical repositories and universal patient medical records offer opportunities for improved collaboration on rare diseases. An improved international computer network would foster collaborative research efforts and would allow much better communication between institutions, so as to share data and help children in the most need.

One obstacle to collaboration is different types of imaging and treatment programmes. Assimilation and comparison of information is challenging when studies have been performed using different protocols at different institutions. By agreeing a set of standard methods on how to perform a certain test, the results will be easier to compare across different institutions. We all make our own images the best quality possible, but sometimes this approach makes imaging studies difficult to compare. National and even international standardisation of protocols will aid study, by reducing the radiation doses used and improving data sharing, which will manifest as improved knowledge and improved patient care

Some of the highest profile paediatric conditions also need reassessment. The medical diagnosis of child abuse is frequently challenged. Parents or caretakers who are correctly or incorrectly accused of harming their child, whether accidentally or deliberately, are sometimes reported in the press, and our imaging, treatments and management systems come under scrutiny. In the intersection of medicine and law, the ability of scientific evidence to prove something beyond all reasonable doubt (the standard required by law) often becomes problematic in demonstrating

what has happened in a particular case. The imaging methods may suffice for medical diagnosis, but may not prove with absolute legal certainty what has happened. Further understanding of fracture mechanics, fracture healing, and helping to rule out other diagnoses will help us make diagnoses with greater certainty, will improve medical management and will aid the courts in their difficult decisions regarding what may or may not have happened in an individual case.

We also need to focus on those diseases which begin in childhood but have life-long consequences. An example is disorders of the hip which begin in childhood, including developmental hip dysplasia, Legg-Perthes disease, slipped capital femoral epiphysis, childhood arthritis and femoral acetabular impingement. These are all disorders of childhood and adolescence which may lead to accelerated degenerative disease as an adult and the eventual need for hip replacement, often at a relatively young adult age. Any treatment that could improve outcome early on in these diseases will have a profound effect on these children during their adult lives. Imaging plays a role in diagnosing these conditions, monitoring their progression and evaluating for complications, and thereby guiding therapy.

On a more generalised basis, disorders such as obesity and diabetes, which begin in childhood, predict adult disease later in life. Imaging does not currently play a significant role in these diseases; however, it may play a role in monitoring overall fat levels or liver fat content over time in such children, or by assessing for early manifestations of coronary artery disease (which is known to start in adolescence). Increasingly, children with congenital heart disease or cancer are surviving into adulthood. For reasons already mentioned, research is needed to minimise radiation dose over the lifetime of such patients. Research is also needed to define how these disease processes may manifest into adulthood, what complications might occur or should be expected, and what role imaging will play in the follow-up of long term survivors of congenital and paediatric diseases.

CONCLUSION

Imaging is paramount for efficient, high quality paediatric medicine. Paediatric imaging continues to advance at a rapid pace. There are many realms in which research is needed to guide this advancement and further improve the healthcare of children.

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"Research is key to the future of paediatric radiology. Investment in well-designed research trials to address today's challenges and questions will improve the healthcare of children tomorrow."

Owen J. Arthurs / Peter J. Strouse



INTERVIEWS
PATIENCE, EXPERIENCE, AND HELP FROM PARENTS: the three key ingredients of successful paediatric imaging

An interview with Fred E. Avni, consultant at the Department of Radiology of Jeanne de Flandre Mother and Child Hospital in Lille, France, and former Chairman of the Department of Radiology of Academic Hospital Erasme in Brussels, Belgium.

European Society of Radiology: What is paediatric imaging? What age are the patients, and how is it different from regular imaging?

Fred Avni: Paediatric imaging includes all the types of imaging investigations (both therapeutic and diagnostic) that are performed in children, from the smallest premature neonate to grown adolescents. You can even extend it to the unborn foetus as most centres for paediatric radiology also perform foetal imaging examinations. This wide range of ages of the patients is the first difference compared to adult imaging.

The same equipment is used as in adults, but it must be adapted and optimised to the size and age of the patient. Another difference to adult radiology concerns the type of diseases that are diagnosed. Any disease can be detected at any age; there are acute or chronic diseases that are typical for each age group, but, especially in the youngest patients, there is a larger proportion of congenital anomalies compared to adults, who tend to suffer more from degenerative diseases

The hospital and technological environment can be quite frightening for children, so patience, experience and help from the parents are necessary in order to obtain good examinations.

Finally, small children are not able to express their symptoms or locate their pain, so you are sort of 'working in the dark' and you have to examine wider areas of the anatomy in order to avoid missing an anomaly.

ESR: Since when has paediatric imaging been a specialty in its own right?

Fred Avni: Paediatric radiology started as a distinct specialty at the end of the 19th century. Historically, the first paediatric radiology centres developed in Graz (Austria), Toronto (Canada) and Boston (USA). The specialty developed further in the 1940s and 1950s in several countries. Interestingly, in the beginning, the

adolescents. times be sedated.

Some older and more conventional imaging techniques are still in use (cystography, chest x-ray, bone x-ray, barium enema and follow-through) and provide helpful information. There is a clear tendency to try to replace irradiating techniques with non-irradiating ones. For instance, conventional radiological cystography has been replaced in many indications by cystosonography, and MR enterography is increasingly replacing small bowel barium follow-through. As mentioned earlier, all these techniques must be

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examinations were sometimes performed by paediatricians, who later completed training in radiology, rather than by radiologists who specialised in paediatric radiology. Progressively, especially since the 1960s, the specialty became a full part of radiology.

ESR: Which imaging modalities are usually used to examine paediatric patients? Does this change depending on the age of the patient?

Fred Avni: All imaging modalities used in adults can or may be used in children. But non-irradiating examinations are preferred to more irradiating ones. Therefore, since the moment it appeared in clinical practice in the late 1970s, ultrasound has been the preferred imaging modality in paediatric radiology. It can be used to image practically all organs or areas, but its use is limited by the presence of air or bones. Its use is also easier in small children than in fatty

The use of magnetic resonance imaging (MRI) is also preferred for a wide range of paediatric applications. The limitations of MRI are the long examinations, which lead to motion artefacts and anxiety in younger children, meaning the patient must some-

Computed tomography (CT) is also used for specific examinations, and the technique provides important information. Newer equipment allows the delivered dose to be significantly reduced. The clinical use of CT should be clearly justified and optimised.

adapted and optimised according to the size of the patient and the indication of the examination.

ESR: Some imaging techniques, like x-ray and CT, use ionising radiation. What risk does this radiation pose to paediatric patients? What kind of safety measures are in place to protect children?

Fred Avni: The risk of ionising radiation has been increasingly debated since the bombing of Hiroshima. Children seem more vulnerable to ionising radiation because of the cumulative effect of radiation and their longer remaining life span. With imaging, a larger proportion of their body is at risk of irradiation. In addition, their organs are constantly developing, with potentially more cells in mitosis, so with a higher susceptibility to radiation.

The real risk is unknown. Still, radiation protection must be applied optimally in order to protect patients according to the ALARA (As Low As Reasonably Achievable) principle. As usual, the first step is to decide whether the imaging examination is justified; if it is justified, it should then be performed. The second step is to verify whether there is no other examination available that would answer the clinical question equally well, but with less irradiation. Finally, if an irradiating technique has to be performed, it should be optimised in terms of radiation protection, using lead protection, collimation, filtration or any other applicable protection method.

ESR: Do general radiologists always use lower radiation doses when imaging children; are there any guidelines to follow?

Fred Avni: Some certainly do; but this question should be asked directly to them. General radiologists do perform a significant number of paediatric examinations and we do not have information on all of these examinations

From many examinations that have been performed in children transferred to our paediatric hospitals that

we receive to review, it seems that unnecessary examinations are performed (especially CT scans and especially in emergency departments) and that not all the examinations are optimised.

Guidelines do exist and should be applied; I presume that all the European and American campaigns regarding radiation safety will increase awareness.

ESR: How aware are parents and relatives of the risks of radiation exposure? How do you address the issue with them?

Fred Avni: The parents and relatives are usually overanxious and under-informed about the real risks of radiation exposure. When performing the examinations, we explain to them what type of examination we are performing and why; we try to reassure them that we are taking all possible measures to reduce the ionising radiation. When possible, we also give them examples that help them understand the real amount of radiation that will be used (e.g. a chest x-ray is the equivalent of 2–3 days of natural radiation).

ESR: Undergoing an imaging examination, especially a long procedure like MRI, can be an uncomfortable and sometimes frightening experience for some children. How can it be made more bearable?

Fred Avni: We try to explain the examination in words and images that the child (if old enough) can understand; we also show him or her the equipment before the examination. Some toys have been built that represent the magnet that the child will lie in; so the patients can play with it prior to the examination. During the examination, a parent can come along and stay close, and the patient can listen to music through earphones. If the anxiety cannot be sufficiently reduced, we might consider some sedation.

ESR: How many imaging exams are performed on paediatric patients in Belgium each year?

Fred Avni: There is no real answer to this question, as a large number are performed by general radiologists in regular hospitals. In a study conducted in 2010, the French Nuclear Safety Authority found that, out of a sample of 100,000 French children, a third had been exposed to at least one examination using ionising radiation. The mean and median effective doses were respectively equal to 0.65 mSv and 0.025 mSv. These values were respectively 5.7 mSv and 1.7 mSv for the children exposed to at least one CT examination – about 1% of the studied population.

ESR: Access to modern imaging equipment is important for paediatric imaging. Are hospitals in Belgium equipped to provide the necessary exams?

Fred Avni: Having the most modern imaging equipment is very important, as it improves performance and usually reduces the length of examinations. Specialised paediatric hospitals tend to have less hi-tech equipment and tend to renew their equipment less frequently. Probably the only exception would be ultrasound machines, which are renewed at a higher rate. Sharing equipment with an adult department could be a solution and could allow easier and more frequent access. In doing this, we can also benefit from the technical experience of engineers who can help us to improve paediatric examinations.

ESR: What has changed in paediatric radiology during your lifetime?

Fred Avni: As for many radiological specialities, paediatric radiology has evolved from a small peripheral specialty of medicine into a large and important specialty that has taken on a significant role in the man*"Patience, experience and help from the parents are necessary in order to obtain good examinations."*

Fred E. Avni

agement of both diagnostic and therapeutic aspects in paediatric patients.

When I started in radiology in the late 1970s, the most sophisticated examinations were intravenous urography (IVU); voiding cystouretrography; barium enema and barium follow-through; x-ray tomography; cisterno-ventriculography; and angiography. Some of them have now disappeared and the use of some others has declined; some are still in use but have been modernised. In general, the philosophy was to obtain the 'nicest image' whereas now we try to get the best diagnostic image, be it nice or not. In addition, we did not 'fear' performing multiple examinations for a single indication, rather than limiting it to the most informative ones. Today, as mentioned, we choose the best and least irradiating technique, following guidelines that are more and more evidence-based.

The introduction of ultrasound has revolutionised the specialty. Paediatric radiology is 'ultrasound first' and the technique should be used whenever possible in children (not forgetting Doppler). CT and MRI are extraordinary techniques and are indispensable for the proper management of many diseases. But they need to be optimised. Their constant development has also improved our performance. In the beginning, we were somehow late in understanding the importance of these new technologies, especially MRI. Fortunately, the situation has rapidly improved. MR urography, MR enterography and MR cholangiopancreatography are developing in children as well.

With time, the workload of paediatric radiology departments has changed, paralleling the evolution of paediatrics and all its specialties. The work up of some diseases like urinary tract infection used to be, and still is, a large part of our practice. They were largely debated without a definite answer and they still are. To do or not do imaging is still the question.

A revolution in our workload came with the increasing use of obstetrical ultrasound in the late 1980s and the discovery in utero of many congenital anomalies. This brought a significant increase in neonatal work-up of many anomalies, especially urinary tract malformations, which has led to the overuse of neonatal voiding cystouretrography. Fortunately it has progressively been reduced. In parallel to the use of obstetrical ultrasound, a new technique appeared in the late 1980s beginning of the 1990s: foetal MR imaging. This also brought another perspective on congenital diseases, their evolution and management. Paediatric radiology had to adapt itself to medical developments, new diseases and new treatments. We had to learn the radiological aspects of new diseases like AIDS for instance; we had to learn how to diagnose and evaluate complications of the chronic diseases occurring at a later age; and we had to learn how to assess normal or malfunctioning transplants, etc.

Overall, we have learned how to image children without harming them, how to reassure them, how to reduce their anxiety or pain, and how to help to cure them.

ESR: Where do you see the next developments in your field?

Fred Avni: New equipment will surely be developed focusing on smaller and more functional imaging. We can expect the rapid development of fusion imaging between different techniques and combined imaging using MRI and positron emission tomography (PET); we will have to see how they can be optimised for children. for contrast will be reduced. ric radiology.



INTERVIEWS

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CT will be less and less irradiating. In addition, the need

Some applications such as elastography are emerging within ultrasound and MRI. We have to assess their role in paediatric imaging. Virtopsy (virtual autopsy) is also a relatively new field of development for paediat-

Finally, the main clinical revolution of recent decades has occurred in genetics. We have to verify how our specialty can integrate information provided by these

advances. We hope that we will be able to cure or significantly improve treatment of diseases in children that are poorly understood today, especially degenerative neurological diseases. New treatments will be developed and interventional imaging will surely be used to bring these new treatments as close to the disease areas as possible.

Interview conducted by Mélisande Rouger, ESR Office

Fred E. Avni is a consultant at the Department of Paediatric Radiology and Head of the Ultrasound Department at Jeanne de Flandre Mother and Child Hospital, University of Lille, France.

He is Associate Professor of Radiology in charge of training graduate and postgraduate students at Brussels Free University. His previous appointments were Chairman of the Radiology Department at the Academic Hospital Erasme (2002-2012) and Chairman of the Radiology Department at the Queen Fabiola Children's University Hospital (1997-2002) in Brussels, Belgium. He specialises in uro-nephropathies, ultrasound, foetal MRI and the development of congenital diseases.

Prof. Avni received a Pioneer Award from the Society of Paediatric Radiology in 2001 and a Gold Medal from the European Society of Paediatric Radiology (ESPR) in 2012. He is an honorary member of the ESPR and the French Society of Radiology. He has authored 150 publications, 30 book chapters, two books and one PhD thesis.



THE CHALLENGES OF **IMAGING CHILDREN:** what one should always keep in mind

An Interview with Alan Daneman, Professor of Radiology at the Department of Medical Imaging, University of Toronto, and Staff Radiologist at The Hospital for Sick Children in Toronto, Canada.

European Society of Radiology: What is paediatric imaging? What age are the patients, and how is it different from regular imaging?

Alan Daneman: Paediatric imaging is a general term used for imaging in children. The general age ranges from the neonatal period until the teenage years. In some specialist paediatric hospitals, the age of patients accepted extends up to 18, but in others the upper age limit may be younger. Paediatric imaging is not only practised in independent children's hospitals, but also in general hospitals, which may or may not have dedicated paediatric units, and it also takes place in private practices. So a significant amount of paediatric imaging occurs outside of paediatric hospitals. Over the past decade, paediatric imaging has also expanded to include imaging of the antenatal foetus. This application has grown since MRI has been used so extensively for foetal evaluation. This allows paediatric radiologists to follow the individual from the stage of the foetus through the neonatal phase and on into childhood.

ESR: Since when has paediatric imaging been a specialty in its own right?

Alan Daneman: Radiologists with a specific interest in paediatric radiology started meeting informally in the early 1950s and this led to the official formation of two large paediatric radiology societies - the Society for Pediatric Radiology in 1959 (based in North America) and the European Society of Paediatric Radiology in 1963. Since then two other major paediatric societies have formed: the Asian and Oceanic Society for Paediatric Radiology (AOSPR) and the Latin American Society for Paediatric Radiology (SLARP). There are also many national societies, e.g. the Australian and New Zealand Society for Paediatric Radiology (ANZSPR). Since the 1970s, there has been the development of dedicated paediatric radiology fellowships in many paediatric hospitals to ensure a high level of intense, dedicated training in imaging practices in paediatrics. These have evolved with further subspecialisation, for example, fellowships in paediatric neuroradiology, pae-

to examine paediatric patients? Does this change depending on the age of the patient? Alan Daneman: All imaging modalities used in adult imaging are used in paediatric imaging. Less invasive imaging modalities are the modalities of choice particularly in young children, but more invasive modalities can be used in young children if need be.

are in place to protect children?



diatric interventional radiology, etc. The societies and the fellowship training programmes have established paediatric radiology as a specialty in its own right.

ESR: Which imaging modalities are usually used

ESR: Some imaging techniques, like x-ray and CT, use ionising radiation. What risk does this radiation pose to paediatric patients? What kind of safety measures

Alan Daneman: Younger children are more sensitive to radiation than older children and therefore modalities not using ionising radiation, e.g. ultrasound and magnetic resonance imaging (MRI), are the preferred choices for this age group. The most effective way of decreasing the amount of radiation exposure to a child is not to use a modality that utilises ionising radiation (see answer below). If a radiation modality has to be used, then the examination should be done with the lowest radiation dose that will provide a diagnostic examination - the As Low As Reasonably Achievable (ALARA) concept.

ESR: Do general radiologists always use lower radiation doses when imaging children; are there any guidelines to follow?

Alan Daneman: Unfortunately general radiologists do not always use lower radiation doses when imaging children. Guidelines have been provided by Image Gently (The Alliance for Radiation Safety in Pediatric Imaging, www.imagegently.org). This is supported by the major paediatric imaging societies listed above. Furthermore, paediatric radiology departments in

large paediatric hospitals have well developed guidelines for radiation protection. These are often accessible on the web and are available to general radiologists.

ESR: How aware are parents and relatives about the risks of radiation exposure? How do you address the issue with them?

Alan Daneman: There has been an increasing awareness of the issues related to radiation exposure to children and over time, parents have become more concerned. However, the risks of any radiation exposure should be weighed against the risks of a disease process if the modality utilising radiation is not used to evaluate the patient. This has to be explained to the parents carefully.

ESR: Undergoing an imaging examination, especially a long procedure like MRI, can be an uncomfortable and sometimes frightening experience for some children. How can it be made more bearable?

Alan Daneman: Young babies can be fed just before the examination and often fall asleep for the duration of the examination. Young children usually require sedation or a general anaesthetic. In older children it is important to explain to them exactly what is going to happen and what is expected from them regarding lying still. There are good videos which are useful to educate these children and their parents regarding the procedure. Once they know what will happen ahead

of time, they are usually better prepared to accept the discomforts. Having a parent in the MRI room with the child is also very helpful to alleviate stress.

ESR: How many imaging exams are performed on paediatric patients in Canada each year?

Alan Daneman: I have been unable to get the figures on how many paediatric imaging examinations are done in Canada in one year. However, in our hospital, which is the largest paediatric hospital in Canada, we do between 135.000 and 140.000 imaging examinations in children each year. The number of MRI examinations continues to increase each year, now accounting for about 14,000 per year. Ultrasound continues to increase but at a slower rate than MRI and we do 25,000 to 30,000 ultrasound exams each year. Nuclear medicine and computed tomography (CT) have decreased over the past decade - CT quite significantly. The number of plain radiographs done each vear remains static.

ESR: Access to modern imaging equipment is important for paediatric imaging. Are hospitals in Canada equipped to provide the necessary exams?

Alan Daneman: In Canada, the major children's hospitals are guite well equipped. However, there is not enough funding to ensure that all equipment throughout a paediatric radiology department is state-of-theart and that enough equipment is available so that waiting times are not too long.

"The development and expansion of ultrasound and MRI have ensured that children have access to imaging modalities that do not use ionising radiation. The role of ultrasound and MRI in paediatric imaging continues to grow, even decades after their introduction in clinical use. In many clinical situations, these two modalities have either completely replaced or led to a marked decreased utilisation of other techniques and modalities that use ionising radiation."

Alan Daneman

ESR: What has changed in paediatric radiology during your lifetime?

Alan Daneman: I have been practicing paediatric radiology for 37 years as a staff radiologist. During this time there have been two major advances in paediatric imaging.

- The development and expansion of ultrasound and MRI have ensured that children have access to imaging modalities that do not use ionising radiation. The role of ultrasound and MRI in paediatric imaging continues to grow even decades after their introduction in clinical use. In many clinical situations, these two modalities have either completely replaced or led to a marked decreased utilisation of other techniques and modalities that use ionising radiation, such as CT, contrast examinations of the GI and GU tracts. Furthermore, US and MRI have enabled radiologists to image certain structures and diseases that previously could either not be imaged at all
- or could only be imaged with much more invasive techniques.
- 2. The development of interventional radiology has enabled safe techniques to be developed which are suitable for paediatric patients. This has led to a decrease in open surgery procedures and often fewer hospital stays.

ESR: Where do you see the next developments in your field?

Alan Daneman: Further developments in US, MRI and interventional radiology will continue to expand the roles of these modalities in paediatric imaging. Nuclear medicine techniques combined with MR will enable more specific diagnoses to be made and will facilitate more accurate follow-up. Intraoperative imaging, particularly with MRI, will facilitate surgical procedures.

Interview conducted by Mélisande Rouger, ESR Office



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Alan Daneman is Professor of Radiology at the Department of Medical Imaging, University of Toronto, and has been a staff radiologist at the Hospital for Sick Children in Toronto for the past 36 years.

He is a member of the Division of Body Imaging at the Hospital for Sick Children and his main interests are neonatal imaging, gastrointestinal imaging in children and imaging of conditions requiring surgical management in children. He has been most involved with sonography and fluoroscopic examinations of the gastrointestinal tract.

He is an Honorary Member of the European Society of Paediatric Radiology (ESPR) and the Latin American Society for Paediatric Radiology (SLARP), as well as the Chilean, Israeli and Hungarian radiological societies, the Brazilian College of Radiology and the Faculty of Radiology, College of Surgeons, Ireland. He has been awarded life membership of the Royal Australian and New Zealand College of Radiologists. He was awarded the Gold Medal of the Brazilian College of Radiology and received two awards from the Society for Pediatric Radiology for his lifetime dedication to education in the discipline of paediatric radiology.

Prof. Daneman has more than 200 publications in peer-reviewed journals to his name and has published a book titled *Pediatric Body CT*. He has been an invited guest lecturer at international and national conferences over 150 times and has given several named lectures.

He has been on the editorial boards of *Pediatric Radiology,* the *Canadian Association of Radiologists Journal,* the *Journal of Clinical Ultrasound,* and the *Journal of the Korean Radiological Society.*



MUCH MORE THAN AN **IMAGING PROCEDURE:** paediatric imaging from a radiographer's point of view

An interview with Vasilis Syrgiamiotis, radiographer at the MRI-CT department of the General Children's Hospital in Athens, Greece.

European Society of Radiology: Radiographers have direct contact with patients and guide them through the whole examination. How is dealing with paediatric patients different to dealing with adults? Are there also differences between different age groups? Vasilis Syrgiamiotis: Radiographers are not only in direct contact with the patient through the whole examination, but they also advise and communicate with the patients before and after the examination, in order to answer the patient's questions.

The imaging procedure for paediatric patients is quite different compared to that of adults. I believe that work with paediatric patients is based on emotional intelligence and various skills the radiographers need to have. Each age group, especially infants and toddlers, has its own complexity; therefore, radiographers need to have expertise in more than just imaging procedures. For example, 99% of infants and toddlers have to be sedated during a magnetic resonance imaging (MRI) or computed tomography (CT) examination, or an interventional procedure; in

such cases, radiographers have to deal with parents as well. Children and teenagers are given a leaflet, which explains the examination procedure; after that, they meet and can talk face to face with the radiographer to clarify any questions. For x-ray examinations, which take place in another department of the hospital, radiographers also participate actively in patient management.

Radiographers always provide necessary information and guidance to parents to help keep the child still, when the child is not cooperative. So we provide optimal information first for parents, and then maintain a calm and friendly atmosphere for the patient during the examination.

ESR: Is your cooperation with radiologists different when imaging children (compared with adult imaging)?

Vasilis Syrgiamiotis: Even if we work as well-oiled machines, we have to be much more careful when examining children due to their complexity. We work

in close cooperation with radiologists to help meet the challenge when using alternative techniques during the examination. Our main aim is to avoid exposing children to unnecessary radiation, because of their higher sensitivity due to their longer remaining life expectancy.

ESR: Some imaging techniques, like x-ray and CT, use ionising radiation. What kind of safety measures are in place to protect children?



ESR: Which imaging modalities are usually used to examine paediatric patients? Does this change depending on the age of the patient?

Vasilis Syrgiamiotis: It is crucial that there is a department policy to avoid ionising radiation. The effect of this policy is that the paediatric patients are mostly examined with ultrasound and MRI. However, when it is imperative to use ionising radiation, techniques such as rational management of exposure factors are used to protect the patient from radiation.

Vasilis Syrgiamiotis: Recent research (Constantarogianni, 2015) indicates that radiographers in Greek paediatric hospitals are very well informed and confident enough to offer information and advice to patients and their relatives about the potential risks stemming from radiation exposure during radiological examinations. Radiographers use dedicated radiation protection measurements in the case of paediatric patients. The personnel of a radiology department in a paediatric hospital have a better patient self-protection score compared to the staff of an adult hospital department. The Council Directive 2013/59 Euratom states in article 56 on optimisation that "member states shall ensure that all doses due to medical exposure for radiodiagnostic, interventional radiology, planning, guiding and verification purposes are kept as low as reasonably achievable." According to this directive, the scientific staff of the paediatric imaging departments intend to optimise imaging protocols and, of course, take into account all the safety equipment that accompanies each procedure.

ESR: Do radiographers always use lower radiation doses when imaging children? Are there any guidelines to follow?

Vasilis Syrgiamiotis: I believe they do when clinically possible. In my opinion, most colleagues aim to optimise radiation dose levels, especially in the case of paediatric patients. Regarding guidelines, the National Dose Reference Levels (NDRL) are defined by the International Commission on Radiological Protection (ICRP) as a form of investigation dose level using easily measured dose quantities.

ESR: How do radiographers keep up to date with current strategies for reducing radiation dose? Do you think radiographers are sufficiently aware of the most effective methods for dose optimisation?

Vasilis Syrgiamiotis: Radiographers should be aware of the latest techniques and protocols. There are a lot of Continuous Professional Development (CPD) activities, courses, webinars, seminars and conferences organised all over the world for radiographers and other members of the radiological team to update their knowledge and awareness of the latest guidelines. According to recent surveys conducted by the European Federation of Radiographer Societies (EFRS), radiographers seem to update their knowledge regularly and they are increasingly involved in research. I can also confirm that radiographers often present strategies and methods in the field of dose optimisation and radiation protection at radiological conferences.

ESR: How aware are parents and relatives about the risks of radiation exposure? How do you address the issue with them?

Vasilis Syrgiamiotis: In my opinion, they are not as aware as they should be. Actually, many clinicians are

not well informed on the matter, as their clinical field is far away from radiology. In many cases, the personnel from other departments are also poorly informed; therefore, radiographers should be the ones who inform patients and their relatives about the potential risks of exposure to ionising radiation. Employees in the radiology department are trained to provide information about the potential risks associated with exposure to ionising radiation and are used to discussing the subject. Indeed, more action is needed. This is indirectly supported by the industry. Leaflets, informative material, and illustrated magazines can certainly raise the awareness of parents.

ESR: Undergoing an imaging examination, especially a long procedure like MRI, can be an uncomfortable and sometimes frightening experience for some children. How can it be made more bearable and how do you handle these situations?

Vasilis Syrgiamiotis: I agree that MRI can be an uncomfortable and sometimes frightening experience even for adults, not only children. I believe that effective communication with the child is key. Raising the parents' awareness of the discomfort arising during an MRI examination may allow easier handling of the situation. Recently in my department, we had the opportunity to add a leaflet with cartoons explaining the MRI procedure in a unique way before the examination to paediatric patients, in addition to real contact. Many systems can now support the examination with music or movies, which can be extremely helpful.

ESR: Can parents and relatives be present and speak to the child during the examination? How do you handle these visitors?

Vasilis Syrgiamiotis: In many cases, the internal rules of each department are what counts. Undoubtedly,

"Work with paediatric patients is based on emotional intelligence and various skills radiographers need to have. Each patient age group has its own complexity; therefore radiographers need expertise in more than just imaging procedures."

Vasilis Syrgiamiotis

most of the time, the presence of parents is key to a successful examination. In my imaging department, we allow parents to be present during the examination. However, we should not forget that maintaining a conversation between parents and children during the examination is likely to result in poor imaging quality. Therefore, talking should be allowed only in specific situations.

ESR: What has changed in paediatric radiography during your lifetime?

Vasilis Syrgiamiotis: I have been working in a paediatric imaging department for seven years. During this time, a tremendous job has been done in the optimisation of paediatric imaging techniques and radiation protection. Projects funded by the European Commission, such as MEDRAPET and EMAN, have already been developed to tackle the issue of ionising radiation. Currently there is an on-going project called European Diagnostic Reference Levels for Paediatric Imaging (PiDRL), coordinated by the ESR and supported by the EC. Faster and improved acquisitions in MRI, digital radiography (DR) and computed radiography (CR) optimise imaging quality. The equipment used in operating rooms (C-Arm) is advanced, which helps to maintain low dose and offers special protective lead for children. Furthermore, we can use special exposure techniques for children, with low exposure times.

ESR: Do radiographers receive special training for paediatric imaging?

Vasilis Syrgiamiotis: This varies across Europe. In some cases, there are a lot of dedicated programmes Interview conducted by Mélisande Rouger, ESR Office

and training opportunities for radiographers working with children. In other cases, there are none. Unfortunately, training and CPD requirements are not harmonised across Europe. Therefore, in 2010, the EC initiated a project to study the implementation of the Medical Exposures Directive (MED) requirements in radiation protection education and training of medical professionals in member states, and the MEDRAPET project to develop recommendations for harmonisation at the EU level. These initiatives resulted in the EC RP175 guideline promoting education standards for all healthcare professionals.

ESR: Where do you see the next developments in your field?

Vasilis Syrgiamiotis: As a radiographer working with MRI and CT in a paediatric department, I feel that the scientific community will try hard to further optimise radiation protection and develop better signalto-noise ratio sequences in MRI. Moreover, in recent years, there has been an increase in studies on the use of ionising radiation in healthcare. Several studies have indicated that healthcare professionals do not have adequate information regarding the level of radiation received by patients. Due to this lack of knowledge, they are not able to adequately and professionally inform patients or relatives. However, radiographers seem to be highly informed and skilled in this area now. I think that a very important issue in my field is justification. Perhaps this issue is just as important as optimisation in all areas of imaging.



INTERVIEWS

Paediatric Imaging



Vasilis Syrgiamiotis is a radiographer at the MRI-CT department at the General Children's Hospital in Athens, Greece. He is also a lab assistant at the Faculty of Health and Welfare Professions of the department of radiographers at the Technological Educational Institute of Athens. He teaches MRI and CT to BSc students, and supervises and coordinates the work of several radiography students in medical imaging.

His research interests include optimisation in paediatric medical imaging and radiation protection. He obtained a Master's degree from the faculty of medicine at the University of Athens, with a thesis focusing on the effect of ionising radiation in children and pregnant women. He is currently a PhD candidate at the same faculty.

Syrgiamiotis has served as a member of the scientific council of the Greek Society of TEI's Medical Radiological Technologists and has sat on the election committee of the European Federation of Radiographer Societies (EFRS). He is currently an EFRS board member for the term 2014–2017, and a member of the Radiographer Scientific Subcommittee of ECR 2016.

He has moderated several national and international scientific sessions and delivered numerous presentations at national and international conferences.









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Prof. Avni received a Pioneer Award from the Society of Paediatric Radiology in 2001 and a Gold Medal from the European Society of Paediatric Radiology (ESPR) in 2012. He is an honorary member of the ESPR and the French Society of Radiology. He has authored 150 publications, 30 book chapters, two books and one PhD thesis.



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Dr. Nanko de Graaf is a Dutch paediatric radiologist. In 2000 he graduated from medical school at the State University of Groningen. After working in Gouda as a hospital doctor in internal medicine, cardiology, and pulmology, he started his radiology residency at Erasmus Medical Center in Rotterdam (chair: Prof. G.P. Krestin). During residency, his focus was interventional radiology. Upon completion of residency, he did a fellowship in paediatric radiology. Afterwards, he completed short observerships in the United Kingdom and United States. He is currently working at Erasmus Medical Center / Sophia Children's Hospital in Rotterdam, the Netherlands.



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include congenital heart disease, paediatric chest pathology, non-accidental trauma, and neonatal cerebral disorders. She has gained broad experience in several radiological methods focusing on paediatric and cardiac MRI, but also PET and ultrasound.

France and in Gothenburg, Sweden and trained as a radiologist in Oslo, Norway. She gained her PhD in 2012, on preclinical animal research, using MRI/contrast-enhanced ultrasound and PET in perfusion and metabolic studies on cerebral birth asphyxia. She is involved in several multicentre studies and research projects and has published several papers in peer-reviewed international journals.

She is a former president of the Norwegian Society of Paediatric Radiology and is currently a Vice President of the Norwegian Society of Radiology.

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Dr. Marina Easty has been a consultant paediatric radiologist since 1998. She trained at the Queen Elizabeth Children's hospital in London and Great Ormond Street Hospital (GOSH) NHS Foundation Trust and was a Consultant at the Royal London Hospital (RLH) when Queen Elizabeth Hospital closed and the hospital was relocated. At the RLH, she was an old-fashioned paediatric radiologist, involved in all modalities including intervention. By 2007, she had left RLH and took up a consultant post at GOSH, where she developed an interest in nuclear medicine and worked both in general paediatric radiology and paediatric nuclear medicine, with a particular interest in hybrid imaging. She is also interested in radiology and nuclear medicine training, radiation protection and radiology errors.



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She is currently completing a PhD on CPD amongst radiographers and the most appropriate mechanisms for ongoing CPD delivery.

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He has published more than 150 peer-reviewed journal articles. 100 review articles or book chapters, and three books related to paediatric imaging. He is chair of the paediatric section of the American Board of Radiology (ABR), an assistant editor of the paediatric section of the American Journal of Roentgenology (AJR); an editorial board member of nine journals; and a reviewer for 25 journals. He is the past President of the New England Roentgen Ray Society (NERRS) and current President of the International Society of Pediatric Thoracic Imaging (ISPTI). Dr. Lee is a sought-after speaker in the field of paediatric thoracic imaging and has served either as a visiting professor or as an invited speaker in more than 20 countries.



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She has lectured at over 150 national and international congresses and supervised higher degrees and diplomas, acting as liaison/host for the European School of Radiology fellowships, and has held grants in excess of £5 million.

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Paediatric Imaging



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Dr. Peter Vock has chaired the Institute of Radiology of the University of Berne from 1989 until 2012. Besides his activities in clinical radiology, his research concentrates on imaging of the chest, CT, and radiation protection. For many years, he has been dedicated to radiation protection on an institutional, professional, national and international level. He has been a member of the Swiss Federal Commission for Radiation Protection for more than twelve years, chairing the Medical Subcommittee for six years. He has also been responsible for radiation protection within the Swiss Society of Radiology and has served the European Society of Radiology as chairman of the Radiation Protection Subcommittee. He has represented the ESR within various radiation protection projects, such as EMAN, MEDRAPET, and PiDRL.

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