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REVIEW

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Imaging of conjoined twins

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Abstract The incidence of conjoined twins is estimated to be around 1 in 250,000 live births. There is a distinct female predominance. In this paper the imaging of conjoined twins both antenatally and postnatally is reviewed, in particular taking into consideration recent advances with multidetector CT. Accurate counselling of parents regarding the likely outcome of the pregnancy and the likelihood of successful separation is dependent on good prenatal imaging with ultrasound and MRI. Planning of postnatal surgical separation is aided by accurate preoperative imaging which, depending on the conjoined area, will encompass many imaging modalities, but often relies heavily on CT scanning.

Keywords Conjoined twins · MRI · CT · Ultrasound

Introduction

Throughout history conjoined twins have appeared in myths and legends. The Greek and Roman god, Janus, had two faces, one young and one old. It has been suggested that centaurs, a combination of horse and man, may have been inspired by ischiopagus or parapagus twins who often have four legs. Conjoined twins have been a source of fascination to the public and medical profession for centuries; stone carvings and statuettes from pre-Christian times are in existence. The earliest record of attempted separation of conjoined twins appears to be from 945 AD in Constantinople (Istanbul) [1]. Known as the Armenian twins, the boys were conjoined at the abdomen (ischiopagus). The separation attempt was performed after one had died aged 30 years, but the other twin succumbed 3 days later. The Biddenden Maids in Kent, UK, born in 1100 AD, were joined at the hips and shoulders and lived for 34 years. After their death, their local church received 20 acres (8 hectares) of land. Subsequently small cakes and biscuits imprinted with the image of the sisters were given to the poor every Easter in remembrance of their generosity. The Biddenden Maids are still honoured by this service over 900 years later.

The most celebrated pair of conjoined twins was Chang and Eng who were born on a riverboat in Siam in 1811. They were joined at the lower chest by a narrow band through which their livers were connected. The Siamese Twins were exhibited during adulthood throughout America and England. They were denied entry into France because officials feared that pregnant women who saw the unusual brothers would bear similarly deformed babies! They lived for 63 years in North Carolina, married sisters and had 22 children between them. Many believe Chang and Eng helped change how society viewed conjoined twins and others with profound physical disabilities. They proved that those who are different can lead normal lives, have jobs, spouses and families. Chang and Eng paved the way for other conjoined twins who have benefited from the acceptance they demanded and generally received from society at large. In addition, none of their offspring was conjoined, indicating the random nature of single ovum twinning. Affected mothers can thus be advised that there is no recognized risk of recurrence.

The frequency of conjoined twins is approximately 1 in 50,000 gestations, but many die in utero, are terminated or stillborn. The true incidence is estimated to be around 1 in 250,000 live births. Like all monozygotic twins, all conjoined twins have the same sex. There is a distinct female predominance with a ratio of 3:1. Conjoined twinning is a random event, unrelated to heredity, maternal

Not all the examinations presented as figures in this article were performed at our institution, but all these patients were referred to us for review. No patient therefore is identifiable from these anonymized images.

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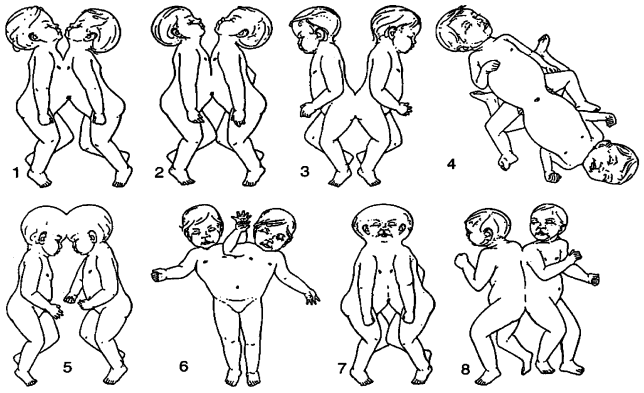
age or parity [2]. According to the fission hypothesis of conjoined twinning, monozygotic or identical twins form when a single fertilized egg splits into two embryos. The phenomenon occurs between the 13th and 15th day after fertilization, when failure to split completely leads to conjoined twins. More recently it has been proposed that these twins may rather result from secondary fusion of two originally separate monovular embryos [2]. Conjoined twins are classified on the basis of the site of union, with the suffix *pagus* meaning fixed or fastened. The classification of conjoined twins, limited to eight types with approximate percentages of frequency, is summarized in Table 1 and illustrated in Fig. 1 [3].

Table 1 Characteristic features of different conjoined twins

Type of fusion	Union		Incidence (%)	Shared structures and associations
	Site of union	Ventral/dorsal		
Thoracopagus	Chest, thorax to umbilicus (one compound or two united hearts)	Ventral (lie face to face)	20–40	Sternum, diaphragm, upper abdomen wall Liver 100% Pericardium 90% Heart 75% (VSD, ASD, tetralogy of Fallot) Upper intestine 50% Biliary tree 25% Common small intestine 50% (typically joins at duodenum, separates at ileum) Exomphalos
Omphalopagus	Umbilicus (may have same union of the trunk as thoracopagus but have two separate hearts)	Ventral	18–33	Heart never fused Liver 80% Terminal ileum and colon 33% (often unite at Meckel's diverticulum) Separate rectum Exomphalos
Pyopagus	Rump (joined at the sacrum)	Dorsal	18–28	Sacrum and coccyx 100% Lower GI tract 25% (single anus, one or two rectums) GU tract 15% (single bladder) Spinal cords usually separate Pelvic bones 100%
Ischiopagus	Hip (from umbilicus to conjoined pelvis)	Ventral (but may lie face to face or end to end)	6–11	Two sacra or two symphysis pubis Lower GI tract 70% GU tract 50% Crossing ureters Tetrapus (four legs), tripus (three), or bipus (two)
Craniopagus	Cranial vault or 'helmet' (joined at any part of the skull except the face or foramen magnum). Vertical (parietal) in majority	Dorsal	2	Skull and meninges 100% Venous sinuses 100% Brains usually separate (some cortical fusion in 33%)
Cephalopagus	Head	Ventral	11	
Rachipagus	Spine	Dorsal	<1	Vertebral anomalies Neural tube defects
Parapagus	Side (ventrolateral fusion); relatively new term (many previously labelled ischiopagus)	Ventral	28	Umbilicus, lower abdomen and pelvis. Single symphysis pubis GU tract 100% Anorectal anomaly and colovesical fistula Anencephaly risk



1. Thoracopagus. 2. Omphalopagus. 3. Pyopagus. 4. Ischiopagus. 5. Craniopagus. 6. Parapagus. 7. Cephalopagus. 8. Rachipagus. (Reproduced with the permission of WB Saunders).

Fig. 1 Types of conjoined twins

We have had the opportunity to examine and treat over 25 pairs of conjoined twins at our institution in the past quarter century. The authors have already written a review paper on the preoperative imaging of conjoined twins [4]. The important aspects of imaging conjoined twins are reviewed in this paper with particular emphasis on prenatal detection and assessment, and in addition recent advances and potential applications of multidetector CT (MDCT) in postnatal evaluation are discussed.

Antenatal diagnosis

Sonography initially and later MRI, particularly in the third trimester when there is less amniotic fluid, but larger fetuses, form the main prenatal imaging modalities. Accurate antenatal assessment allows the parents to be counselled as to the probable outcome of the pregnancy and the likelihood of successful postnatal separation. Prenatal diagnosis of conjoined twins is important for optimum obstetric management, including decisions regarding timing and method of delivery to minimize maternal and fetal mortality. Vaginal deliveries have been complicated by dystocia, fetal and maternal morbidity, and would not now be recommended when the likelihood of the twins' postnatal survival is good.

CT has been performed antenatally in the assessment of conjoined twins but the high radiation burden, to the young mother in addition to the twins, precludes its routine use, such that CT has been superseded by prenatal MRI.

Sonography

Antenatal US is capable of diagnosing a conjoined twin pregnancy as early as 12 weeks gestation [5]. Transvaginal US may also aid early diagnosis [6]. Diagnosis of conjoined twins may be straightforward when fusion of fetal parts is obvious (Fig. 2). In addition, the possibility of conjoined twins should be suspected in a twin

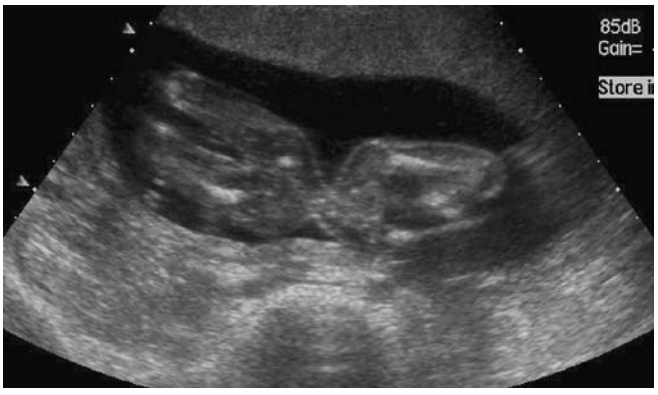


Fig. 2 Antenatal US image showing fused abdomens in omphalopagus twins (courtesy of Prof. Nick Fisk)

pregnancy with a single placenta and no visible separating amniotic membrane. Conversely, observation of two placentas or an amniotic membrane excludes conjoining [2]. The US findings depend, of course, on the area of fusion, but include inseparable fetal bodies, unvarying relative positioning of the two fetuses, both heads persistently at the same level to each other, bibreech or bicephalic presentations, fewer limbs than expected, and a single umbilical cord with more than three vessels. Most conjoined twins face each other and are fused ventrally, resulting in hyperextension of their cervical spines [2]. Polyhydramnios complicates up to 50% of conjoined twin pregnancies, compared to about 10% of normal twin pregnancies. Detailed US assessment at around 20 weeks gestation should be able to define the site and extent of the conjoined area and provide a reasonable evaluation of which viscera are, and are not, shared.

Fetal ECHO assessment of the heart needs to be detailed in all patients as there is an increased incidence of congenital heart disease in conjoined twins overall. A particularly detailed cardiac assessment is vital in thoracopagus twins as a shared heart is seldom compatible with life and usually an indication for termination of the pregnancy. Hearts can be confirmed as separate when they are seen to be anatomically separate or when the heart rates are different. It has been the experience of many authors that fetal ECHO underestimates the severity of cardiac anomalies [2]. The thoracopagus heart typically is six-chambered. Due to the abnormal cardiac anatomy and function, an increased nuchal translucency and subcutaneous oedema have been noted in thoracopagus twins in particular [7]. Communications at the atrial and ventricular levels can often be well defined with US. However, the great vessel relationships and atrial morphology are often difficult to determine [7]. Prenatal US does, nevertheless, provide an opportunity to accurately define the morphological connections between the various chambers of the hearts. That is important as the state of the cardiac conjunction in particular is the fundamental pointer to eventual outcome. Prenatal ECHO is made easier by amniotic fluid, particularly polyhydramnios in later pregnancy, acting as an acoustic window, and the lack of lung aeration permits US evaluation from different approaches.

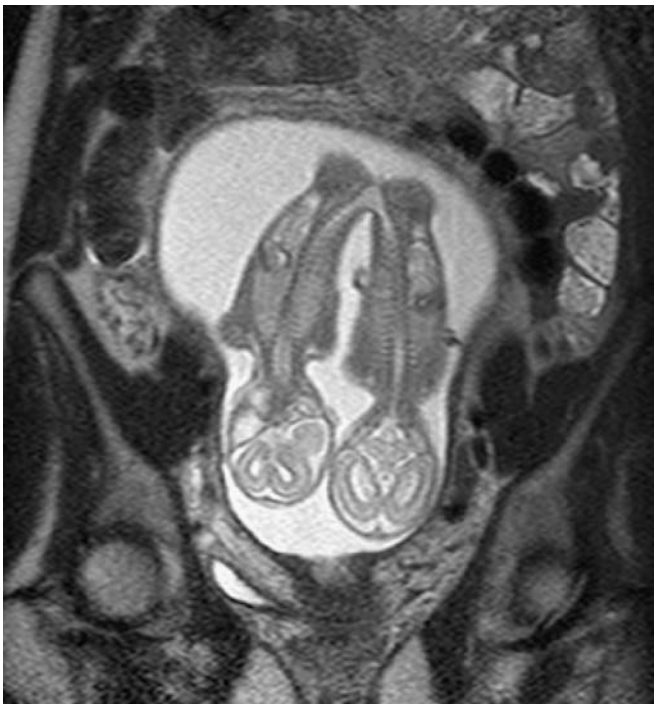


Fig. 3 Prenatal T2-W MRI showing fusion of the distal thecal sacs in pyopagus twinning

Postnatally, the small precordial window, fused thorax and lung aeration hinder thorough ECHO evaluation.

Recently 3D US imaging has been advocated as a new tool to demonstrate the extent of fusion in conjoined twins. 3D US may add anatomical information and thus improve the accuracy of classification of individual twins. In some cases the category of conjoined twins is suspected by 2D scanning, but can be better appreciated on 3D US. It would seem sensible however to postpone attempts at detailed 3D US until after 14 weeks gestation when better views of the anatomy can be obtained [5]. Although 3D US may add more information regarding the extent of the union in utero, it is questionable whether these findings affect management prenatally.

MRI

In later pregnancy, especially if there is maternal obesity or oligohydramnios, MRI will be superior to US for overall fetal assessment. MRI, with its ability to differentiate soft tissues, provides an excellent alternative technique. Ultrafast T2-weighted (T2-W) sequences of short duration, such as the single-shot fast spin-echo sequence, allow minimal image degradation by fetal motion and high-quality images of fetal organs without the need for fetal or maternal sedation [8]. The larger field of view of MRI permits better evaluation of the spatial relationships of anatomical anomalies or between normal structures (Fig. 3). Additional 3D MRI models may be useful in specific circumstances [9]. Bladder exstrophy, which is often difficult to appreciate on antenatal US, has been shown prenatally by MRI [10]. There are no reported adverse biological effects of MRI, which is thus far considered a safe imaging modality in pregnancy. Prenatal MRI has also recently been advocated to be of value when one twin is confirmed to be very unlikely to survive, such that planning for an extrauterine intrapartum (EXIT) procedure and immediate separation is facilitated [11, 12].

Fig. 4 Plain radiography. **a** Ischiopagus twins joined end to end. **b** Scanogram from a CT of parapagus twins with three legs (tripus). **c** Omphalopagus twins joined from the thorax to the umbilicus. It is worth noting that both twins need not be examined when only one twin needs radiography ▶

Postnatal imaging

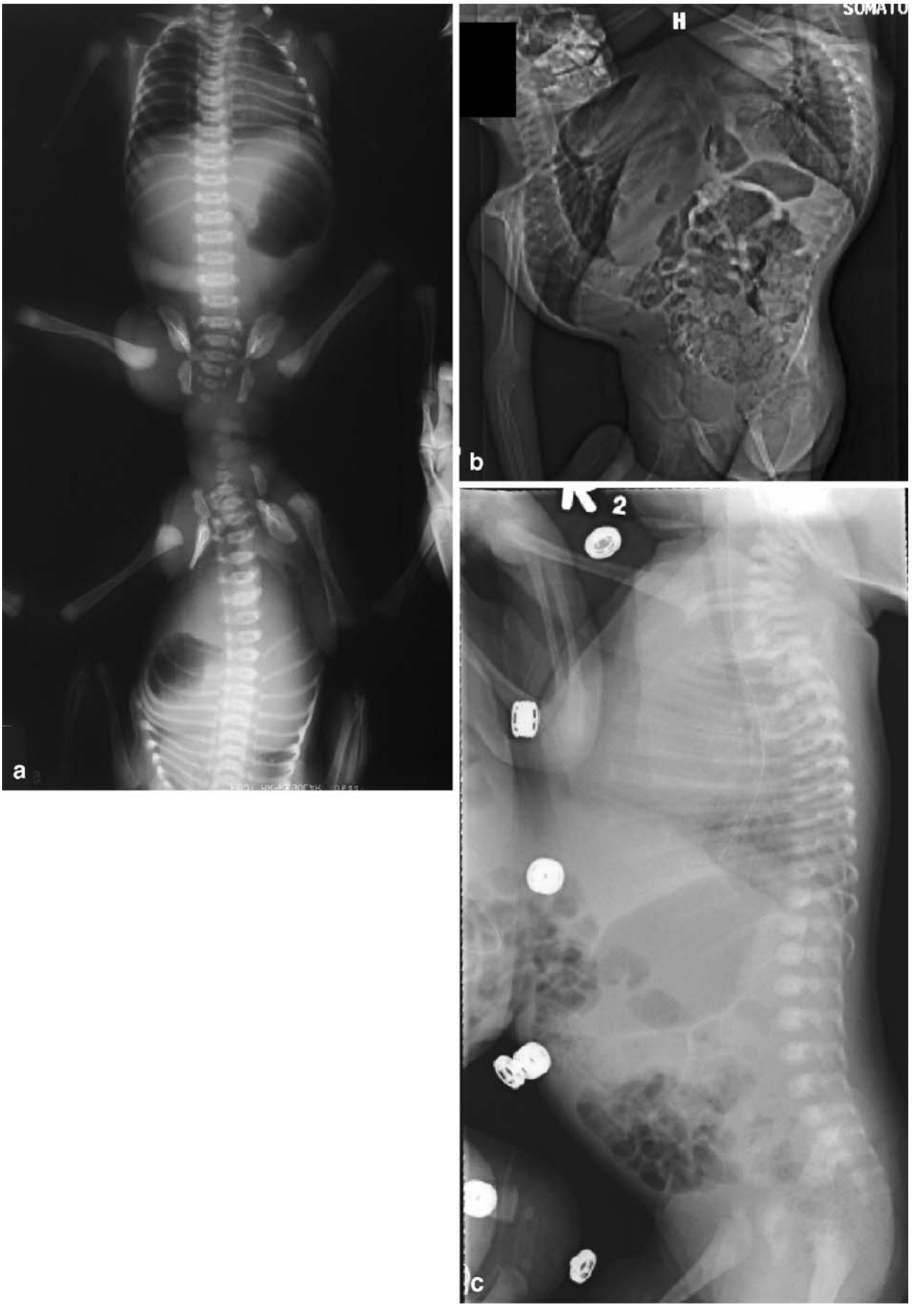
The choice of imaging study will depend to some degree on the site of fusion. Nevertheless, all twins inevitably have chest and abdominal radiography for an overall general assessment, and partly to help health professionals understand the extent of the conjoined area. Unexpected diaphragmatic hernia or vertebral anomalies can thus be detected early (Fig. 4).

Sedation and anaesthesia

The anaesthetic management of conjoined twins has been reported elsewhere and is beyond the scope of this article [13]. It is noteworthy, however, that the response of conjoined twins to medications can be unpredictable when shunts or unexpected cross-circulation cause immediate mixing of their blood. Drugs administered to one twin may have unexpected effects on the other with responses often differing in each twin. Complete colour-coded sets of separate equipment and medication can be made available for each twin [14]. Endotracheal intubation may be difficult due to acute flexion or rotation of the necks. Limited neck mobility, the position of the faces, or frontal conjunction in craniopagus twins may complicate airway access. There may be an increased tendency in these twins to laryngeal irritability with an increased risk of laryngospasm. The merits of laryngeal masks and fibre-optic intubation are discussed elsewhere [15–17]. CT and occasionally MRI can be performed in newborns with a mere ‘feed and wrap’ approach without the need for sedation. Older infants usually need general anaesthesia for CT or MRI, administered separately by two anaesthetic teams.

Sonography

All neonates should have routine cerebral US, and probably spinal US also, as baseline investigations. In addition, abdominal US to assess the liver, to document the presence of two spleens, gallbladders, biliary systems, kidneys and bladders, is necessary. Detailed Doppler studies to evaluate the great vessels in the abdomen and


a
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2
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hepatic venous drainage should also be performed, but midline abdominal conjunction may make accurate Doppler assessment unreliable. Meticulous labelling of the images, ensuring the correct twin is consistently noted to be on the same side, needs to be adhered to at all times (Fig. 5). ECHO is mandatory for every twin due to the high frequency of congenital heart disease in all types of conjoined twins. 3D ECHO has been advocated postnatally as it may facilitate an easier understanding of the cardiac connections, and can thus be helpful to parents [18].

CT

Radiologists avoid CT where possible in infancy due to the high radiation burden. Conjoined twins, even when stable and asymptomatic, are a well-justified exception. Due to the high spatial resolution and speed in particular of multidetector CT (MDCT), MDCT is the best overall modality for evaluating conjoined twins in the postnatal setting.

Contrast enhancement is mandatory to assess the vascular anatomy and so prior cannulation to minimize the stress of the procedure is advisable. Intravenous contrast agents for CT are calculated per kilogram of combined weight. We have successfully used 2 ml/kg of 300 mg iodine /ml contrast agent for the combined weight, but lower doses are likely to be adequate on MDCT. Delayed images give very useful information regarding excretion by both twins' kidneys, the ureteric anatomy, and the location and number of bladders (Fig. 6). Separate studies examining each twin's vascular anatomy on different days are recommended.

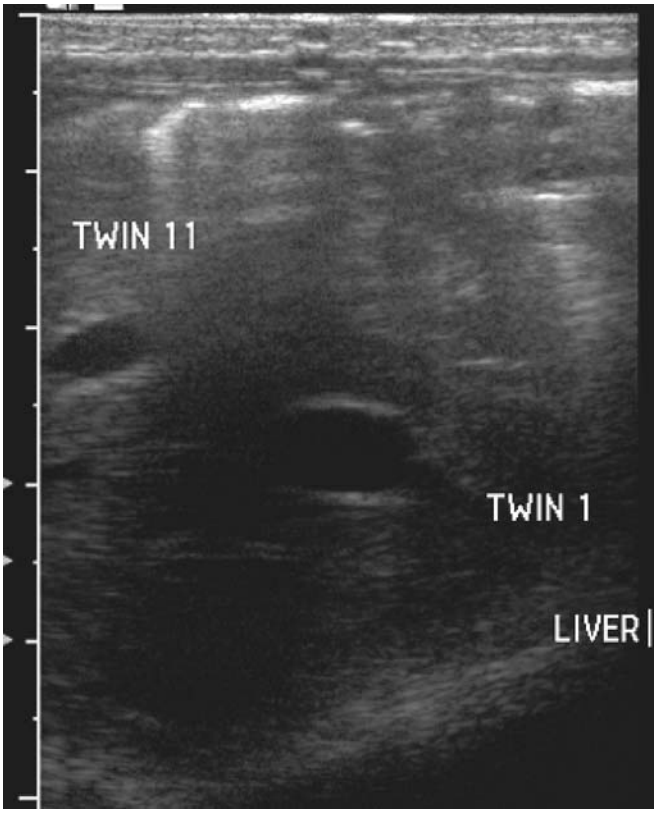

Fig. 5 Abdominal US showing two gallbladders in twins fused at the abdomen. Note that correct and consistent labelling of each twin's images must be adhered to

MRI

MRI has an increasing role in the postnatal evaluation of conjoined twins, particularly those joined at the head or thorax. MR has the capability of producing 3D reconstructed images in any direction with much improved resolution and tissue characterization than 3D US. MR is the optimum examination to assess for any cortical fusion in craniopagus twins. Although the ultrafast sequences, such as single shot T2-W images, have advantages in the constantly moving fetus, more conventional T1-W, T2-W and T1-W images after IV gadolinium administration are utilized in the postnatal imaging of conjoined twins. Intracardiac anatomy, great vessel anatomy and blood flow, and ventricular wall motion can all be accurately assessed in thoracopagus cases. Many complex fusions will inevitably get both MRI and MDCT. These modalities are often complementary with MRI showing soft tissue anatomy to best effect and CT detailing complex bony anatomy in pelvic conjunction. MRI with MR cholangiopancreatography (MRCP) is likely to offer the best hope of assessing the biliary anatomy [9, 11].

Contrast studies of the gastrointestinal and genitourinary tracts

Urological abnormalities are confined to those in whom the pelvis is joined: ischiopagus, parapagus or pyopagus twins. Most twins share four kidneys and two bladders, occasionally with one ureter crossing from the contralateral twin to the other. The bladders are usually side by side but they may be sagittally placed, which presents a reconstructive challenge [19]. CT scanning usually provides sufficient information regarding the upper renal tracts and bladders. Detailed urethral anatomy and possible fistulas require retrograde contrast medium examinations.

Abdominal conjunction often involves fusion of parts of the intestine (Table 1). In addition, there is always a common peritoneal cavity. Even in the absence of fused bowel, when contrast medium is given to one twin the bowel loops may be freely mobile across the 'midline' and be seen to project into the peritoneal cavity of the other twin during fluoroscopy (Fig. 7). Pelvic conjunction leads to complex fusion and anomalies in the anorectal region. These require an individual approach with contrast studies (enemas, loopograms and cystograms) of the distal bowel and bladder. Sharing of undivided intestine may involve the lumen, only the muscularis or the serosa [20]. Consequently, although performed in many cases, in practice, contrast studies provide limited information and often, particularly due to overlapping bowel loops, may not be as revealing as hoped [10, 12].

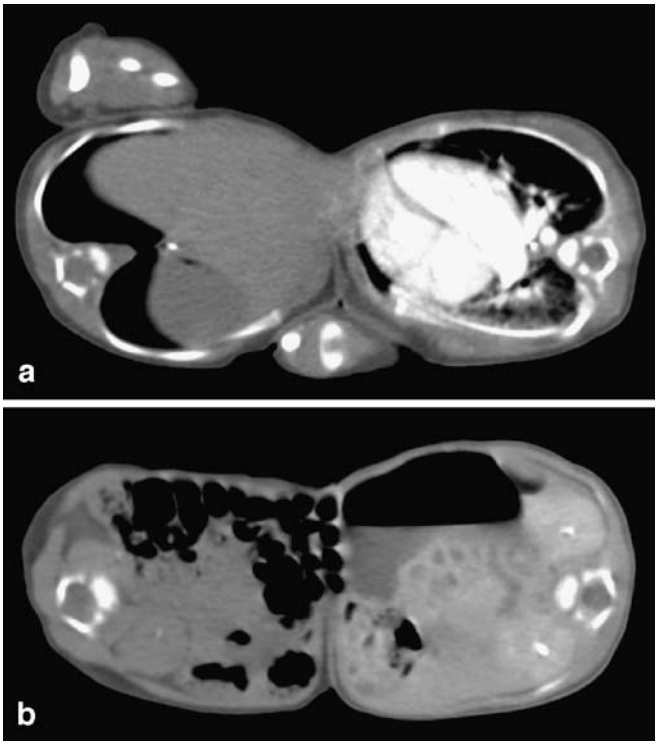

Fig. 6 CT. **a** Contrast-enhanced CT of one twin fused at the thorax shows enhancement only in that twin's heart and aorta indicating separate hearts, i.e. omphalopagus twinning. **b** Delayed images more inferiorly show four contrast-enhanced kidneys (although only a small amount of contrast medium in the contralateral twin's upper kidney is seen here)

Nuclear medicine

The kidneys, albeit ectopic or too few in number, usually function normally in conjoined twins. Cross-sectional imaging can usually clarify the genitourinary anatomy without the need for isotope renography studies. There is an increased frequency of pelviureteric or ureterovesical obstruction in these twins, but this is seldom of major importance prior to separation. Although hepatobiliary agents have been used to demonstrate separate biliary drainage, these are seldom necessary in practice. When two gallbladders are definitely seen in thoracopagus or omphalopagus twins the likelihood of separate biliary systems is high [21]. When twins share the liver or have shared hepatic veins or other large shunts, then mixing of blood precludes the use of a hepatobiliary radiopharmaceutical. Despite detailed preoperative imaging, complex biliary anatomy may only become apparent at the time of surgery. MRCP may have a particular role to play in this area in the future. Individual twins raise unique individual dilemmas and nuclear medicine studies may nevertheless be useful in specific circumstances.

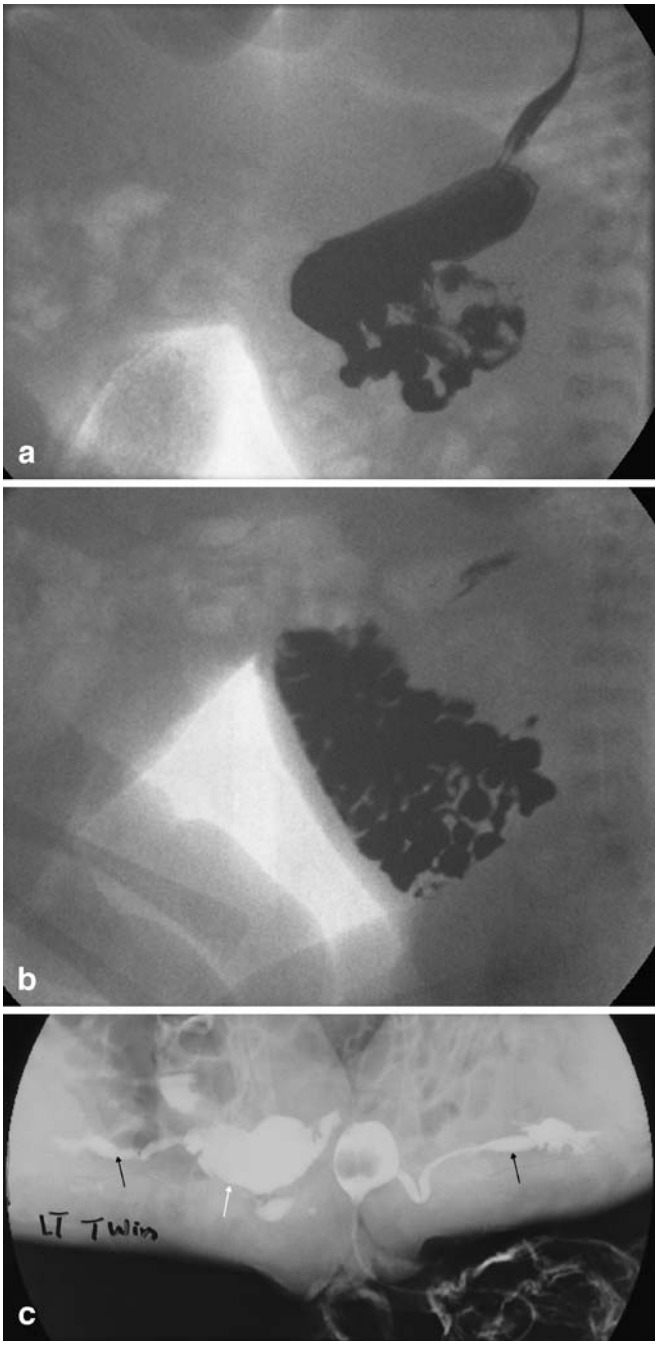

Fig. 7 Contrast studies. **a**, **b** Images from an upper gastrointestinal study in omphalopagus twins. Although no fused small bowel was present in this case, because all children with abdominal conjunction have a common peritoneal cavity, bowel loops will be seen to cross the midline into the other twin's abdomen during fluoroscopy. **c** Retrograde contrast study in ischiopagus twins with a common anus. A distended balloon was placed in the rectum. There is flow of contrast medium into a distended rectosigmoid (*white arrow*). Reflux also occurred into one ureter and collecting system of each twin (*black arrows*)

Postnatal imaging: radiological dilemmas and considerations

Thoracopagus

Approximately 80–90% of thoracopagus twins are not candidates for surgical separation due to the complex nature of cardiac conjunction, and associated cardiovascular anomalies. Thoracopagus twins with ventricular conjunction have never been successfully separated with both twins surviving. Only a single case of separation of twins with conjoined atria has been reported [11]. The cardiac anatomy can be usefully categorized into four types: (1) separate hearts and pericardium, (2) separate hearts but common pericardium, (3) fused atria and separate ventricles, and (4) fused atria and ventricles [18]. Conjoined hearts may possess more than six chambers, some of which may be rudimentary.

Liver anatomy can be best appreciated after IV contrast enhancement, either at CT or MRI. Additional coronal or 3D reconstructions will provide further information. Demonstration of separate hepatic venous drainage into the IVC and right atrium of each twin is important as absent or severely anomalous hepatic venous drainage in one twin is incompatible with survival after surgery.

Omphalopagus

As there is no mixing of blood in the cardiac chambers, the liver can be assessed by CT or MRI with contrast medium injection into one twin. Imaging is commenced soon after injection, during the early arterial phase of enhancement. Non-enhancement of one heart confirms no cardiac conjunction (Fig. 8). The liver parenchyma receiving the hepatic arterial supply from the injected twin enhances and the site of fusion is marked by irregular lobules, which can also be seen during surgical separation (Fig. 9) [4].

The stomach and proximal small bowel are usually separate, and each twin has a rectum. In up to one-third of omphalopagus twins, the terminal ileum and colon are shared, and may also have a dual blood supply. These cases may benefit from angiography prior to separation; such angiographic studies should now be possible with the improved resolution of newer MDCT scanners. There is usually no union of the genitourinary tract.

Ischiopagus

Ischiopagus twins lie face to face or end to end with the vertebral column in a straight line. The twins are termed tetrapus (four), tripus (three) or bipus (two) according to the number of legs attached to the conjoined pelvis. The most common arrangement is four legs. The spinal columns are usually separate.

Pelvic conjunction leads to complex urogenital and orthopaedic anatomy and requires detailed cross-sectional imaging (Fig. 10). MDCT in the early arterial phase will show the skeletal anatomy of both twins (there is seldom a need for non-contrast CT scanning), in addition to the pelvic arterial supply of one or both twins depending on their cross-circulation. In addition, delayed images are necessary to show excretion of contrast medium into the ureters and bladder(s). The kidneys usually function normally, but are often malrotated or ectopic in location. When two bladders are present they lie side by side in a collateral position or they may lie in a sagittal midline location with one bladder draining into the other. The ureters frequently cross over and insert into a contralateral bladder such that they will need to be re-routed during separation.

Partial urethral duplication is possible but a single urethral orifice is typical. The distal gastrointestinal tract is often shared, with anorectal agenesis and rectovesical fistula. Contrast studies are necessary to delineate distal

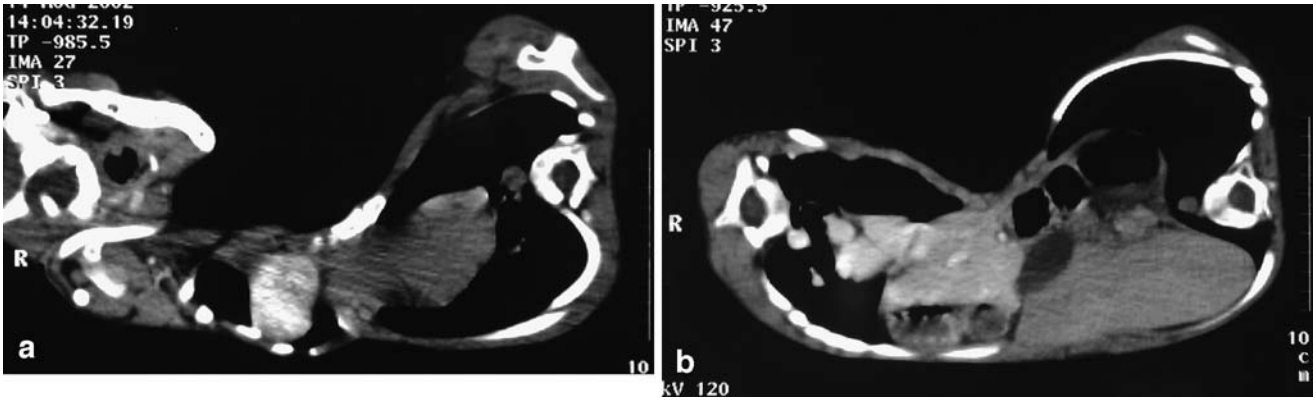

Fig. 8 Contrast-enhanced CT. **a** The right twin shows no cardiac conjunction as enhancement is limited to the right twin. Note omphalopagus twins need not lie at precisely the same level. The unenhanced viscus of the left twin on this image is the upper liver (the left twin was more superiorly positioned). **b** More inferiorly, the enhancing liver of the right twin is seen adjacent to the unenhanced liver of the left twin, with the gallbladder of the left twin also seen interposed between the livers. Another gallbladder was seen on lower sections, indicating separate biliary systems

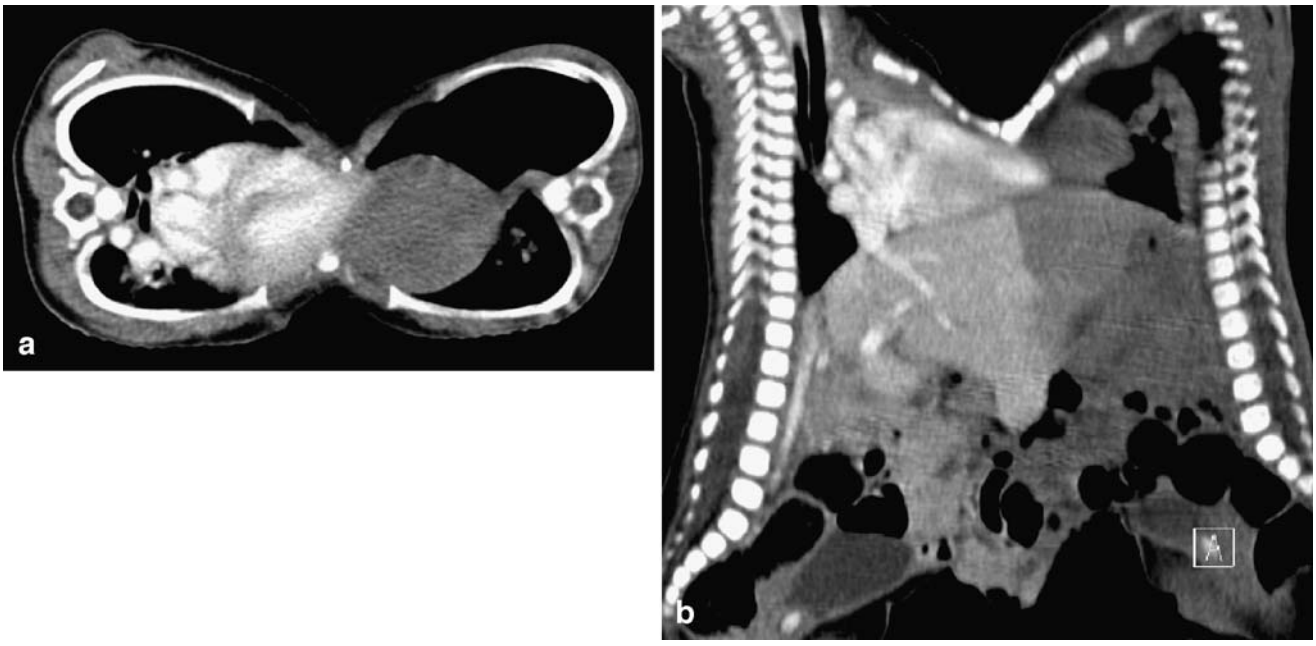

Fig. 9 CT of omphalopagus twins. Axial CT (**a**) with sagittal reconstruction (**b**) after contrast enhancement of the right twin, show no cardiac or hepatic fusion

bowel anatomy. Urogenital sinus or cloaca require contrast studies and multiplanar MRI to clarify the presence and relationship of any separate uterus, vagina, cervix and ovaries. Both CT and MRI can show separate sacra (Fig. 11). In boys there is an increased incidence of undescended testes.

Pyopagus

Pyopagus twins are joined dorsally, sharing the sacrococcygeal and perineal regions. They face away from each other. There is an increased incidence of vertebral anomalies, including hemivertebrae, hemisacral agenesis and thoracic anomalies [22]. Mirror image anomalies do not appear to occur with different regions of the spines

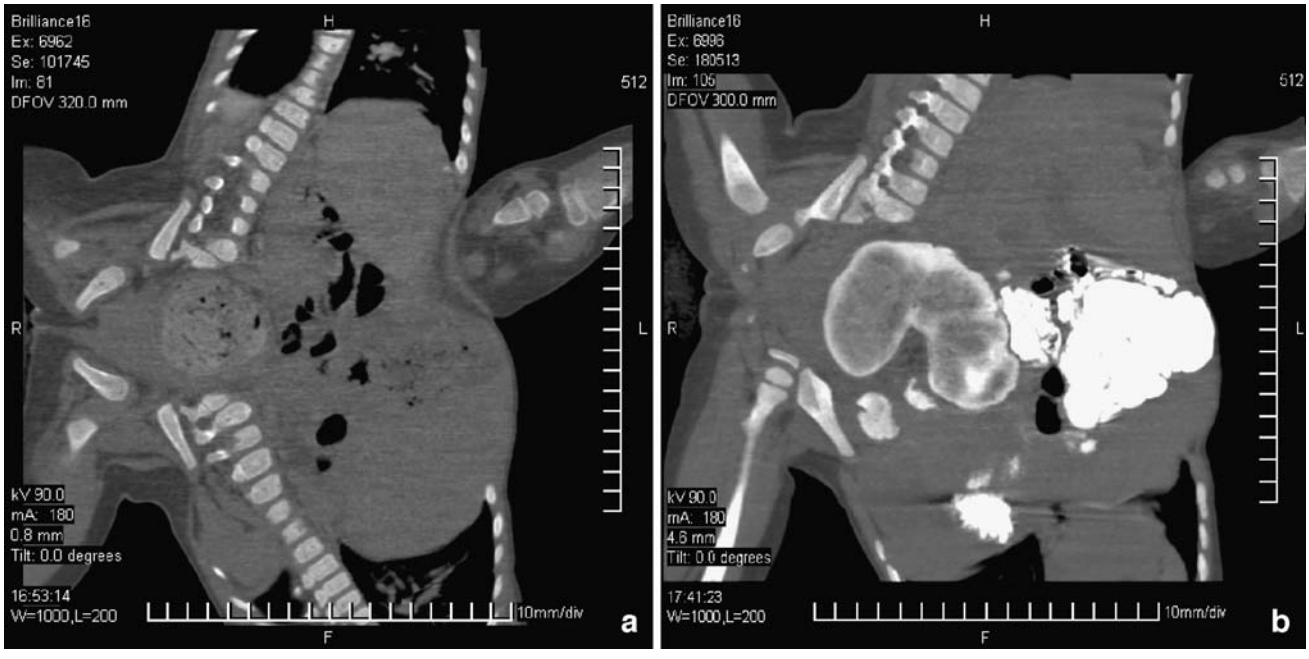

Fig. 10 Ischiopagus twins. **a** Coronal reconstructed CT image shows three legs (tripus). The left leg here was rudimentary with each twin having one normal hip joint. **b** Similar orientation on a different day showing contrast medium in a common distended rectum and a normal hip inferiorly

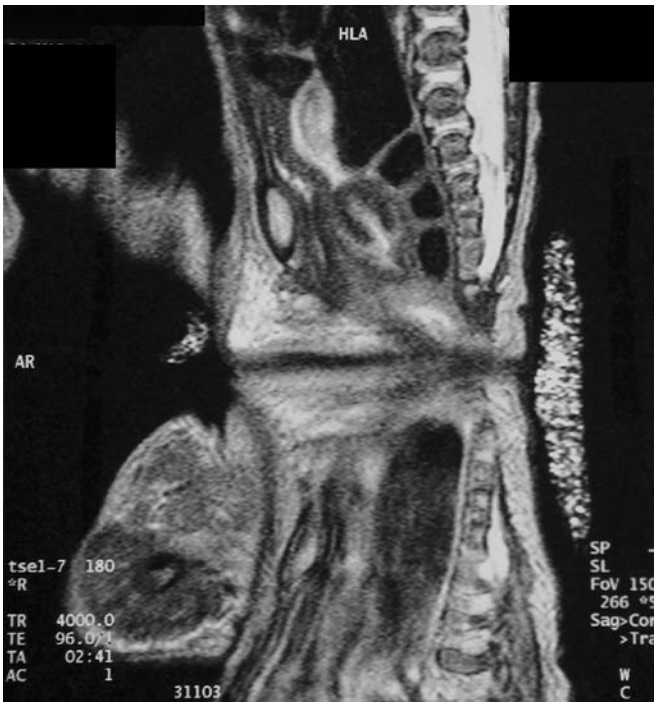

Fig. 11 Sagittal T2-W MRI showing ischiopagus twins with fusion of the perineum, but their sacra and spinal canals are separate

involved in each twin. Although the pelvic conjunction is fundamentally different than in ischiopagus twinning, the types are similar insofar as numerous other associated orthopaedic anomalies have been reported in association with pelvic conjunction, such as hip subluxation or dislocation, congenital vertical talus, talipes equinovarus, Sprengel shoulder and scoliosis. There can also be a variable degree of spinal fusion. Clinical examination or electromyography may suggest spinal cord fusion, easily detected with spinal MRI. A 'Y'-shaped appearance to the fused lower cords on MRI has been reported in this setting [23]. Although there may be only one anus and rectum, the remainder of the intestines are usually separate. The upper bodies are not fused and there are four arms and four legs.

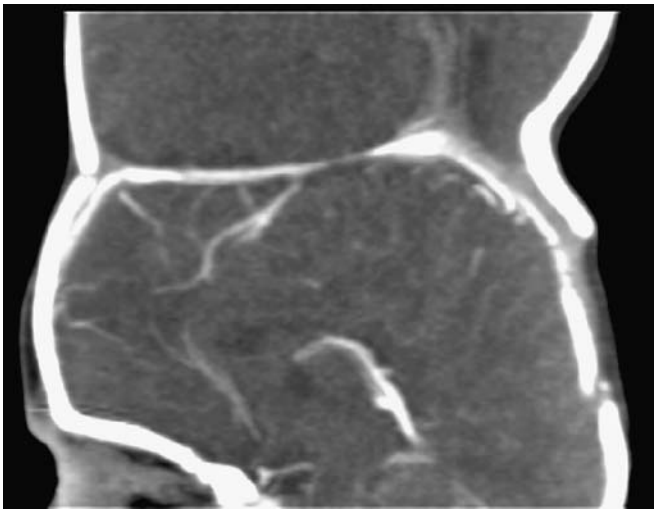

Fig. 12 Contrast-enhanced CT showing vertical (parietal) craniopagus twins sharing the superior sagittal sinus

Craniopagus

O'Connell [24] initially classified craniopagus as partial or total. Partial was defined as limited surface area involvement with intact crania or cranial defects. The abnormality extends no deeper than the leptomeninges. Total craniopagus was defined as sharing an extensive surface area with widely connected cranial cavities. Later four general subtypes were adapted: frontal, parietal, temporoparietal and occipital [25]. Frontal craniopagus twins face each other, occipital twins face away from each other. Parietal craniopagus occurs when twins fuse at the vertex, and this alignment is often referred to as vertical craniopagus (Fig. 12). Vertical craniopagus twins lie in the same long axis, but varying degrees of rotation may result in the twins facing in the same or opposite directions [25].

The majority of complete craniopagus twins have only dura separating their brains. Portions of the cerebellum may be seen to herniate into the other twin's posterior cranial fossa. Craniopagus twins may show fused or interdigitated regions of cortex, easily identified with MRI. Shared brain tissue impacts significantly on postoperative morbidity and when extensive may preclude surgery. Bucholz et al. [26] reported that all cases with profound neurological deficits had cerebral connections prior to surgery.

Craniopagus twins develop a common shared superior sagittal, or 'circumferential' sinus which is often the most challenging aspect of twin separation. It is a common factor in twin morbidity and mortality. The twin who receives the common sinus at surgical separation inevitably has the lower morbidity and mortality, avoiding venous hypertension and cortical infarctions. Endovascular balloon test occlusion may predict the need for revascularization as an option for managing the shared venous drainage [27]. Staged surgical separations, with gradual re-routing of the shared blood supply, have been used successfully in some craniopagi [25].

Solid anatomical models, generated by the patients' own imaging data, have been used in particular with craniopagus twins [28]. Translating complex medical image data into solid objects allows true tactile interaction with patient anatomy in 3D. These models can be useful in many stages of the separation process including surgical planning and communication with the families for informed consent [28].

Cephalopagus

Cephalopagus twins often have a fused thorax in addition to a fused head. The single fused head may have two faces (janiceps) facing away from each other; one face may be rudimentary. Other variants exist and have been examined post-mortem with MDCT [1]. These twins are terminated or die in utero. They are nonviable (Fig. 13).

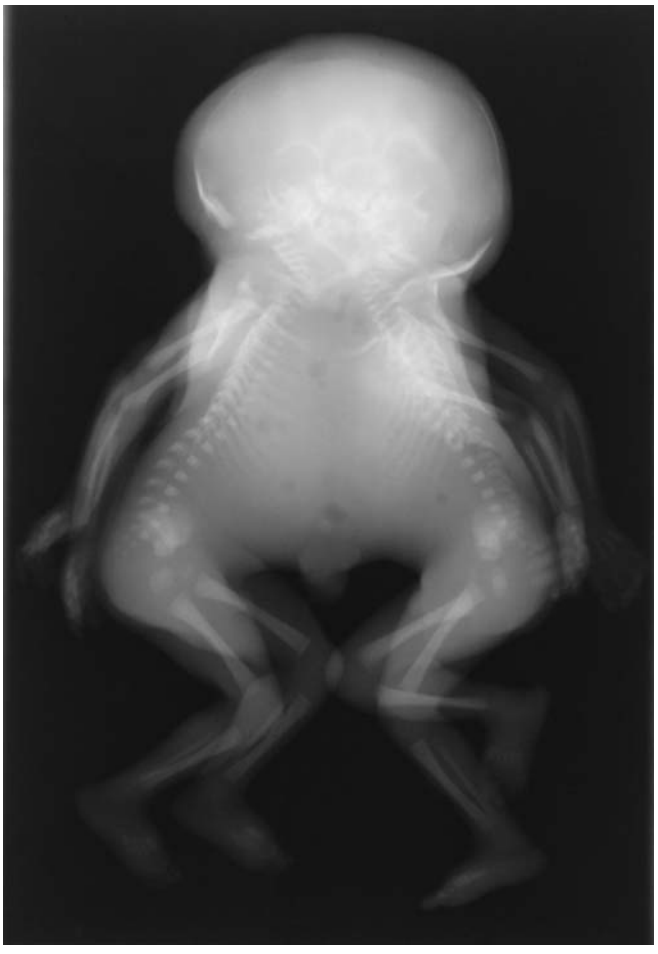

Fig. 13 Post-mortem radiograph of nonviable cephalopagus twins

Rachipagus

Rachipagus twins, joined at the vertebral column mainly by the vertebral arches, are the rarest of all conjoined twins [29]. Out of a review of 1,200 conjoined twins, Spencer [30] could find only one example of typical rachipagus twinning. Those reported twins had dorsal union of the entire head and trunk. The most common site of union in rachipagus is in the dorsal midline. It appears that most rachipagus twins, however, are parasitic in type with in utero demise of one of the twins, typically with persistence of a conjoined limb with additional supernumerary structures.

Parapagus

Parapagus twins are joined side to side with extensive ventrolateral fusion. The conjoined pelvis has a single symphysis pubis and one or two sacra. The thorax may be fused. When the fusion involves only the pelvis and abdomen and the thoraces are separate, the twins are termed *dithoracic*.

Thoracic union requires detailed cardiac assessment. Ventricular cardiac union is reported in parapagus twins, but that has not been our experience, although many of our parapagus twins have had a common pericardium [18]. Defects of lateralization including right and left atrial isomerism and mirror imagery are known to be particularly common in parapagus twins.

The imaging studies performed will be similar for other twins fused at the abdomen and pelvis. Liver fusion requires assessment with contrast-enhanced CT. Contrast studies of the distal bowel are performed to evaluate for bowel sharing, anorectal malformation and colovesical fistula. Complex urogenital abnormalities, unique to each twin, need careful evaluation with US, CT/MRI and often retrograde contrast studies. The vascular anatomy is also complex, but MDCT should now obviate the need for catheter angiography in many cases, notably when the vascular supply of the shared leg needs to be demonstrated. Orthopaedic anomalies are again frequent. There are two, three or four arms and two or three legs.

In conclusion, male twins in general have a worse outcome. Despite genetic identity, conjoined twins often have discordant anomalies. These anomalies generally occur more often on the right side. When laterality defects are present, they occur on the right-sided twin in approximately 86% of parapagus twins, and 71% of thoracopagus twins [4]. The propensity to laterality defects depends on the orientation of the conjoined twins [31]. Twins not joined obliquely, such as those fused at the head (craniopagi) or pelvis (ischiopagi) show few laterality defects. The site of fusion is also prone to anomalous development, e.g. exomphalos at the site of the conjoined umbilicus. Some anomalies are intriguing and pose questions on the origins of conjoined twinning [3]. For example, diaphragmatic defects and cleft lip/palate are most common in cephalopagus and parapagus diprosopus (two faces); anencephaly is frequent in parapagus diprosopus, moderately so in cephalopagus, but rare in craniopagus. Other anomalies are surprising and unexpected, such as coarctation in one twin, and it is a truism in the investigation of conjoined twins to be always on the lookout for unsuspected abnormalities [32].

The site of conjunction determines the radiological approach. Cardiac assessment is mandatory in all conjoined twins, particularly those with thoracic fusion. MDCT allows better vascular assessment in neonates and infants than was previously possible, obviating the need for conventional angiography in many cases. Bone windows facilitate assessment of complex bony anatomy and skeletal fusion, aided by reformatted images in the sagittal and coronal planes.

The occurrence of conjoined twins raises moral and ethical dilemmas that are beyond the scope of this article. Intrusive media coverage further complicates an already difficult clinical situation. Each set of twins poses unique challenges. Although high-quality radiology can predict most anomalies preoperatively, due to the rarity of these cases not all their complex anatomy may be conclusively demonstrated prior to separation surgery. Nevertheless, suffice it to say here that appropriate and high-quality diagnostic imaging is fundamental to surgical planning,

and better preoperative radiology is undoubtedly a factor in the reported improved outcome in many recently reported cases.

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