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Conjoined twins – role of imaging and recent advances

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Abstract

Introduction: Conjoined twins are identical twins with fused bodies, joined in utero. They are rare complications of monozygotic twinning. The purpose of this study is to describe the various types of conjoined twins, the role of imaging and recent advances aiding in their management. **Material and methods:** This was a twin institutional study involving 3 cases of conjoined twins diagnosed over a period of 6 years from 2010 to 2015. All the 3 cases were identified antenatally by ultrasound. Only one case was further evaluated by MRI. **Results:** Three cases of conjoined twins (cephalopagus, thoracopagus and omphalopagus) were accurately diagnosed on antenatal ultrasound. After detailed counseling of the parents and obtaining written consent, all the three cases of pregnancy were terminated. Delivery of the viable conjoined twins was achieved without any complications to the mothers, and all the three conjoined twins died after a few minutes. **Conclusion:** Ultrasound enables an early and accurate diagnosis of conjoined twins, which is vital for obstetric management. MRI is reserved for better tissue characterization. Termination of pregnancy when opted, should be done at an early stage as later stages are fraught with problems. Recent advances, such as 3D printing, may aid in surgical pre-planning, thereby enabling successful surgical separation of conjoined twins.

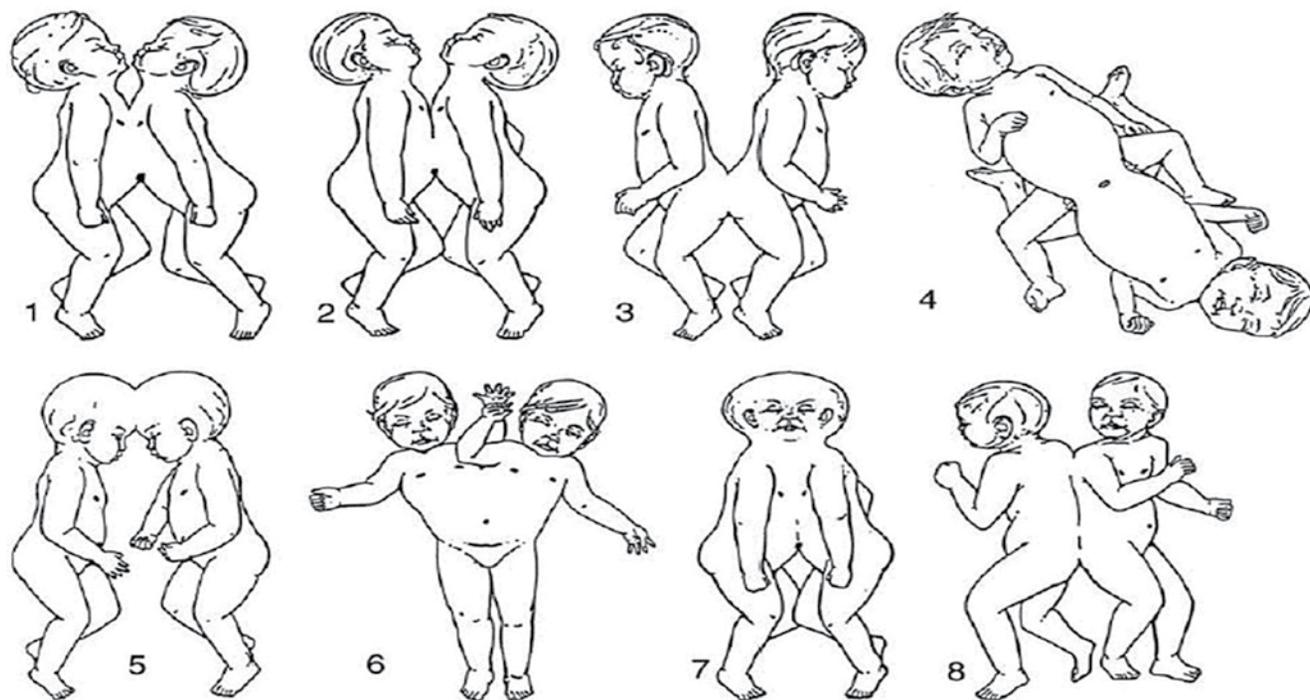
Introduction

Conjoined twins are a rare and extreme form of monozygotic twinning. The incidence of conjoined twins falls in the range of 1:50,000 to 1:100,000 live births, with 40–60% being stillborn and about 35% of the live births not surviving beyond 24 hours. They are classified based on the site of fusion (Fig. 1). Conjoined twins are popularly known as Siamese twins, named after the birth place of the original Siamese twins born in 1811 in Siam (Thailand). Both of these twins were males and lived for nearly 62 years and

fathered 22 children⁽¹⁾. Imaging plays a crucial role in the antenatal and postnatal life of conjoined twins, with the initial modality for screening being ultrasound followed by other modalities such as computed tomography (CT) and/or magnetic resonance imaging (MRI)⁽²⁾.

Material and methods

This study involved 3 cases of conjoined twins diagnosed over a period of 6 years from 2010 to 2015. Institutional



1. Thoracopagus, 2. Omphalopagus, 3. Pygopagus, 4. Ischiopagus, 5. Craniopagus, 6. Parapagus, 7. Cephalopagus, 8. Rachipagus

Fig. 1. Schematic diagram showing the various types of conjoined twins

ethical committee clearance was obtained prior to the commencement of the study. All the 3 cases were evaluated and diagnosed antenatally by experienced radiologists using ultrasound machines – Philips IU-22 and GE Voluson P8, with curved array transducers (1–5 MHz). Each fetus of the 3 twin cases was individually evaluated from the cranium to the foot. The site of fusion was noted. The number of vital organs, such as heart and liver were noted. The spine was evaluated for the presence of congenital defects or fusion. The pelvis was evaluated for the bladder, genitals and for fusion abnormalities. The number of limbs was noted. Color Doppler was applied for identifying cardiac defects or malformations and for evaluating the hepatobiliary system. The placentas were evaluated for the number and any abnormality. Detailed histories of the patients and their husbands were taken regarding exposure to drugs, environmental teratogenic agents and prior medication or surgery for infertility, as well as for a family history of twins.

The first was a case of cephalopagus twins (Fig. 2) of 28 weeks gestational age diagnosed in a 29-year-old primigravida. Ultrasound (Fig. 3) revealed conjoined twins fused from the top of the head down to the umbilicus with a single large face with hypotelorism, two unfused thoracic spines, fused thorax with two hearts, eight limbs, and two separated pelvises.

The second case was a pair of thoracopagus twins (Fig. 4) referred from an outside diagnostic center with a fetal



Fig. 2. Postnatal images of the cephalopagus twins

gestational age of 25 weeks in a 26-year-old woman. Ultrasound examination revealed conjoined twins, fused face to face from the upper thorax to the upper part of the abdomen. The heads were fully developed and the twins had a common sternum, diaphragm and upper abdominal wall. A single shared heart with two atria and ventricles was noted. A fused liver with a single biliary system was noted. The pelvises and urinary tracts were separate. Each of the twins had 2 pairs of limbs.

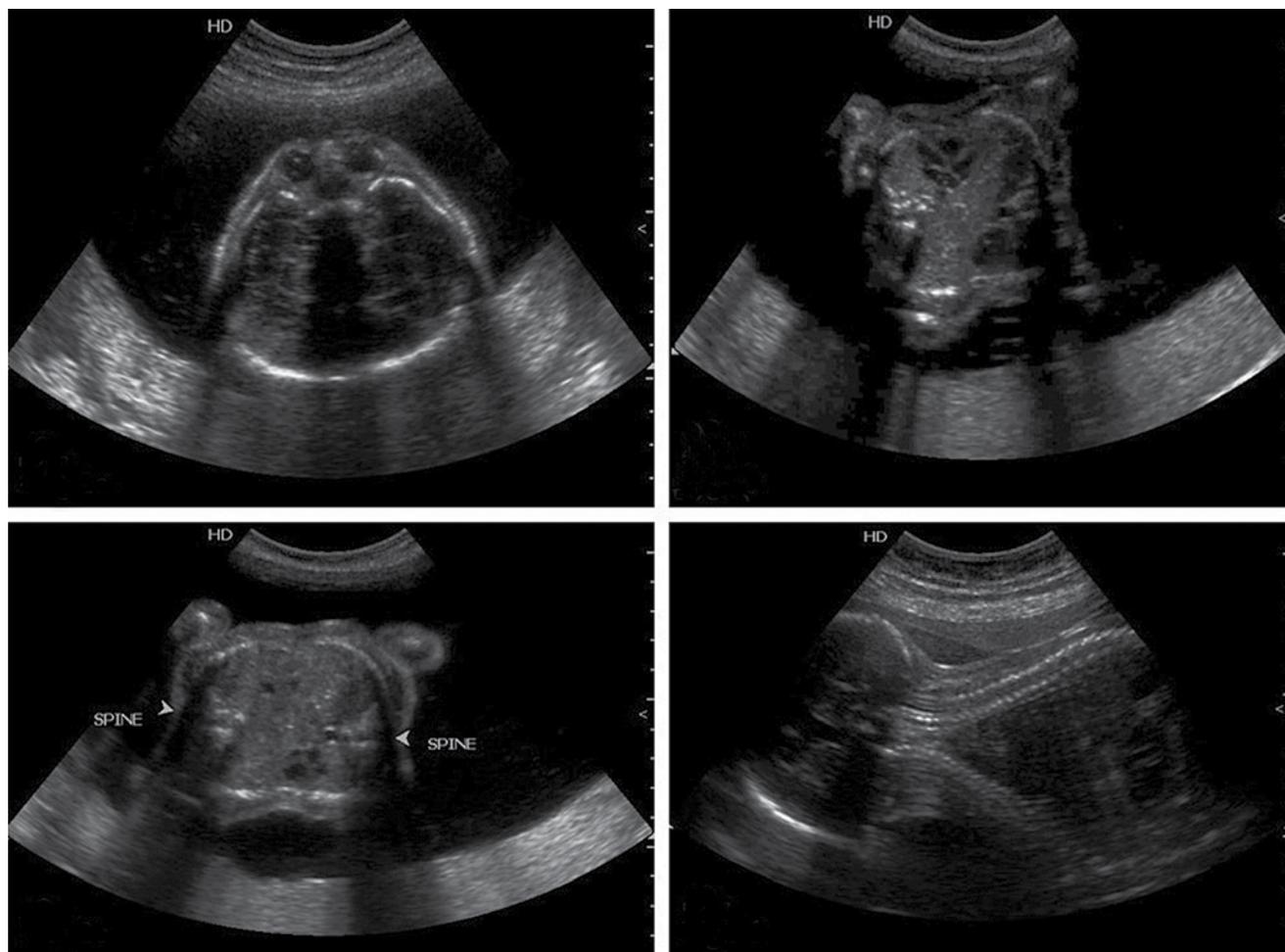


Fig. 3. Ultrasound images of a case of cephalopagus twins



Fig. 4. Postnatal image of the thoracopagus twins

The last case was an 18-week gestational age omphalopagus fetus. On ultrasound (Fig. 5 A, 4 B), the twins were fused from the lower chest to the umbilicus, with separate heads and chests (lungs and hearts). Two separate fully developed spines were noted. In the abdomen, 2 stomachs and a common shared liver were noted. Due to the small gestational age, a detailed evaluation of the biliary system and intestines could not be done. The twins had separate pelvises, urinary systems and 2 pairs of limbs each. After explaining the malformations to the parents and obtaining written consent, MRI examination (Fig. 5 C) was done for further evaluation. Due to the limited experience in fetal MRI combined with the fact that twins were only 18 weeks of gestational age, not much information was obtained.

All of the 3 cases had no prior 1st trimester dating scans done.

Results

After detailed counselling and obtaining written consent from all the parents, both the cephalopagus and thoracopagus twin pregnancies were terminated by cesarean section, while the omphalopagus twin pregnancy was ter-



Fig. 5. Ultrasound (A, B) and coronal T2-weighted MR image (C), showing a case of omphalopagus twins

minated by medication (vaginal misoprostol) (Fig. 6). The post-operative periods in all the cases were uneventful and the set of conjoined twins died a few minutes post-delivery.

Discussion

Conjoined twins are an extreme and rare form of monozygotic twinning with an incidence of 1 in 50,000 to 1 in 100,000 live births. Most of these conjoined twins are females, with a female to male ratio of 3:1⁽²⁾. The two most common theories behind the origin of conjoined twins are the “fission theory” and “fusion theory”, with the latter

gaining more acceptance. According to the fission theory, the embryonic disc undergoes an incomplete separation about 13–15 days post fertilization. While according to the fusion theory, two separate mono-ovulatory embryonic discs undergo a secondary association. Conjoined twins are classified based on their site of fusion – thoracopagus (thorax), omphalopagus (abdomen), pyopagus (sacrum), ischiopagus (pelvis), craniopagus (skull), cephalopagus (face) and rachipagus (back) (Tab. 1)^(3,4).

Antenatal assessment

An antenatal diagnosis of conjoined twins can be made on ultrasound in the 1st trimester. Features favoring a diagnosis of conjoined twins on antenatal ultrasound include – fetuses lying in the same constant position with heads and body parts at the same level persistently; inseparable body and skin contours; fetuses facing each other with hyperflexion of cervical spines; fewer limbs than expected; sharing of organs; and a single umbilical cord with more than 3 vessels^(5,6).

Technical factors related to imaging

Sedation and anesthesia

As conjoined twins often have vascular shunts and cross circulation, their response to anesthesia can be unpredictable due to the possibility of circulatory admixing. Imaging of neonate or premature infants is often possible in the absence of anesthesia. The twins are fed shortly before the procedure and are immobilized using swaddling, vacuum bean bags and other devices. In older twins, two separate anesthetists are used, one for each twin, and general anesthesia is administered after intubating them simultaneously. The preferred route of anesthetic drug administration is mostly intravenous. Intravenous doses of premedications and anesthetic agents for the total body weight of both twins are halved and then divided into two parts, to be ad-



Fig. 6. Postnatal images of the omphalopagus twins

Types	Definitions
<i>Thoracopagus</i>	Twins joined face-to-face from the upper thorax to the upper part of the abdomen; the heart is always involved
<i>Omphalopagus</i>	The fusion includes the umbilicus region, frequently at the lower thorax, but never the heart
<i>Pyopagus</i>	Twins fused dorsally sharing the perineal and sacrococcygeal areas; only one anus but two rectums
<i>Ischiopagus</i>	The union usually includes the lower abdomen and duplicated fused pelvic bones; external genitalia and anus are always involved
<i>Craniopagus</i>	Twins joined by the skull, sharing the meninges but rarely the brain surface; the face and trunk are not involved
<i>Parapagus</i>	Twins joined laterally, regularly share the pelvis. Varieties of parapagus conjoined twins are parapagus dithoracic (separated thoraces), parapagus dicephalus (one trunk two separate heads), and parapagus diprosopus (one trunk, one head, and two faces)
<i>Cephalopagus</i>	There are two faces and twins are joined from the top of the head to the umbilicus
<i>Rachipagus</i>	Twins fused dorsally; the defect may involve the dorsolumbar vertebral column and rarely the cervical vertebrae and the occipital bone

Tab. 1. Classification of conjoined twins

ministered one for each twin. Radionuclide tracers such as technetium-99m sulfur colloid, radiolabeled albumin or red blood cells and inhaled $^{15}\text{O}_2$ are useful for assessing the amount of vascular shunting, cross circulation and exchange of blood volume between the twins^(7,8).

Contrast media

The dose of contrast media administered intravenously is calculated per kilogram of total body weight of both twins. The non-ionic iodine-based contrast is administered separately for each twin. Experts advice a maximum dose of 3 mL/kg of combined weight for these studies. Contrast-enhanced MRI is done using gadolinium at a dose of 0.2 mL/kg (0.1 mmol/kg)^(9,10).

Imaging of various types of conjoined twins

Thoracopagus

These types of conjoined twins are fused from the upper thorax to the umbilicus and share a common sternum, diaphragm, and upper abdominal wall. Nearly 90% of them share a common pericardial sac with some degree of cardiac fusion. The overall prognosis and success of separation is determined by the severity of cardiac fusion^(11,12). After echocardiography, the twins are further evaluated by cardiac angiography/ magnetic resonance angiography. Most of the time, the liver is fused with a common

biliary system. Better information can be obtained by multiplanar MRI and sagittal CT. The biliary system can be adequately evaluated by 99m- technetium hepatobiliary iminodiacetic acid ($^{99\text{m}}\text{Tc-HIDA}$) scan. The visualization of two separate gall bladders along with the excretion of contrast into separate small intestines indicates an independent extrahepatic biliary system for each twin. In indeterminate cases, an intra-operative cholangiography or MR cholangiopancreatography may be useful. These twins have separate large intestines, pelvises and urinary tracts, along with separate sets of limbs^(12,13).

Omphalopagus

These twins are united anteriorly in the umbilical region. The heart is unaffected; however, they may share the common pericardium. The majority (80%) have the common liver. The stomach and proximal small intestines are separate; however, 30% have fusion of the small intestine at the level of Meckel diverticulum in the distal ileum. Distally, the colon separates and each twin has a separate rectum and both the twins have separate sets of limbs, pelvises and urinary systems^(3,13).

Pyopagus

These twins are fused posteriorly, facing away from each other with a common sacrococcyx and perineum. Although the sacral vertebrae are often fused, these twins often have separate spinal cords. They have the common anus with two separate rectums, and separate intestines proximally. These twins have two sets of limbs. If spinal cord fusion is suspected clinically and on electromyography, further assessment by angiography and MRI is needed to assess viability of separation^(7,11,13).

Ischiopagus

Ischiopagus twins are joined from the level of the umbilicus extending caudally leading to a large fused pelvis. They have separate spinal cords and lie facing each other or end to end with the vertebral column in a straight line^(11,14). Depending on the anatomy of the shared pelvis, these twins may be classified as: *tetrapus (four)*, *tripus (three)*, or *bipus (two)* depending on the number of legs attached to the fused pelvis. The most common are the tetrapus twins. These conjoined twins require and in-depth evaluation from an orthopedic and urologic point of view⁽¹⁵⁾. These twins have complex and varied genitourinary anomalies. Initial assessment of the genitourinary system is by ultrasound, followed by a detailed study of the urinary tract and bony pelvis by multi-detector computed tomography (MDCT). The detailed ureteral pathways can be studied by using intravenous pyelography (IVP). These twins usually have a single external urethral orifice, which may be studied by cystoscopy or micturating cystourethrogram (MCUG). Renal abnormalities can be adequately assessed using functional isotope renography. MRI and ultrasound

can be used to evaluate the genitals^(13,16). When surgical separation is considered in twins with shared genitalia, one of twins will not receive any of the genitalia and will undergo a gender change. The lower digestive tract is often shared up to the anus, with anal atresia and colovesical fistula being frequent⁽¹⁷⁾.

Craniopagus

These twins are fused in any location of the skull other than the face or foramen magnum. In more than 60% of the cases, the fusion is vertical and parietal in location. The cranium, meninges and dural venous sinuses are most often shared, with the brains being separate but connected by a neural bridge of tissue. These twins also have separate bodies and limbs⁽¹⁸⁾.

Parapagus

Parapagus twins lie on either side with anterolateral fusion having a common umbilicus, abdomen and pelvis with a single pelvis and one or two sacra. The twins are called *dithoracic*, if they have separate thoraces and fusion involves only the abdomen and pelvis. The term *dicephalic* is used when the heads are separate and the trunks are fused. These twins may have between two to four upper limbs, while two or three lower limbs. They share the pelvis and have anal atresia, with colovesical fistula being a common finding. Using a combination of ultrasound, intravenous urography (IVU), cystography and nuclear scans, genitourinary anomalies can be adequately evaluated. For vascular anatomical studies, a combination of aortography with selective arteriography may be helpful. An adequate assessment of the musculoskeletal system may be carried out by MRI and/or CT⁽¹⁹⁾.

Cephalopagus

These twins are rare and are fused from the vertex to the umbilicus. They usually have two faces on either side of the fused head, with one face being rudimentary. They have the common heart and liver with separate lower abdomen and pelvis. Each twin has two pairs of limbs. Separation of these twins is usually not possible.

Rachipagus

Rachipagus twins are very rare, and are fused posteriorly face away from each other, with the fusion terminating above the sacrum. The occiput and vertebral column are variably involved^(11,19).

Associated anomalies

Anomalies that have been described in conjoined twins have been elaborated in Tab. 2⁽²⁰⁾.

Thoracic anomalies	Dextrocardia (in thoracopagus and dicephalic parapagus twins) Congenital diaphragmatic hernia Anomalous pulmonary venous drainage
Gastrointestinal anomalies	Meckel's diverticulum Bowel atresia Anomalous hepatic venous drainage
Genitourinary anomalies	Duplex system Renal dysplasia Pelviureteric junction obstruction Vesicoureteric junction obstruction
Musculoskeletal anomalies	Congenital dislocation of hip Clubfeet Vertical tali Scoliosis

Tab. 2. Anomalies associated with conjoined twins

Role of 3-dimensional (3D) printing in the management of conjoined twins

3D printing traces its history to the 1980s when Chuck Hull, a US inventor, patented a stereolithography apparatus. 3D printing is the term used for the designing and generation of physical models. Further advancements were made in the 1990s by Mr and Mrs. Crump and others like Andy Christensen. However, its entry into clinical use was much slower due to the limitations of computing and imaging power. Jonathan (Jay) Morris and colleagues, in 2008 at Mayo Clinic, Rochester, successfully performed a highly complex surgical separation of conjoined twins using pre-surgical 3D models. The 3D models were the exact size of the vital organs to be separated and helped the surgeons understand the spatial relationships. Currently, 3D printing plays 3 major roles, namely surgical planning, patient specific simulations and education. Advantages offered by 3D printed models include pre-surgical planning, reduced time in the operating room, reduced patient morbidity, huge cost savings, better patient outcomes as well as education of everyone in the team from scrub techs to medical trainees and all potential learners⁽²¹⁾.

A similar complex surgical separation of a pair of conjoined twins connected from the chest all the way down to the pelvis was carried out successfully by a group of surgeons at Texas Children's Hospital, Houston, in 2015 using a 3D printed model. Post-anatomical computerized segmentation, color-coded output was exported for 3D printing, which vividly displayed the twins' heart, lungs, stomach and kidneys, and all sites in which they were connected (Fig. 7). As per the experts from the center, a combination of volumetric CT, 3D modelling and 3D printing are to become a standard part of preparation for surgical separation of conjoined twins, despite barriers remaining to its adoption⁽²²⁾.

Conclusion

Antenatal imaging by ultrasound plays a vital role in the early intrauterine diagnosis of conjoined twins. Successful

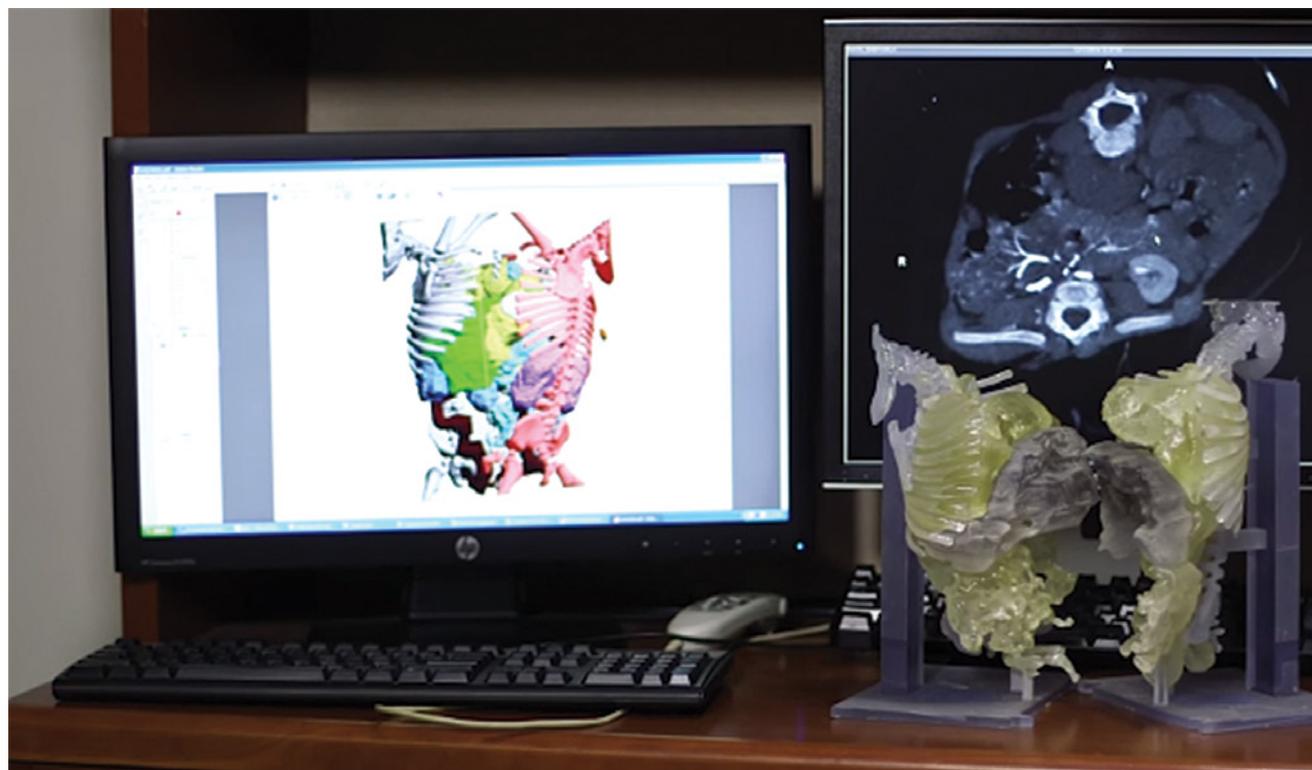


Fig. 7. 3D color segmentation of CT data with the 3D printed model

separation of conjoined twins requires the joint effort of a team comprising various specialists, including radiologists, obstetricians and pediatric subspecialized surgeons. The surgical team requires to be aware of the latest developments and available tools, such as 3D printing which will not only help them in pre-surgical planning but may also help to reduce morbidity and complications. In developing countries, like India, where there are limitations in expertise and technology, detailed counseling of the parents is required to explain long-term outcomes. When

termination of pregnancy is opted, it should be done at an early stage as later stages are fraught with problems.

Conflict of interest

Authors do not report any financial or personal connections with other persons or organizations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

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