Diagnostic Techniques and Criteria for First-Trimester Conjoined Twin Documentation: A Review of the Literature Illustrated by Three Recent Cases

Leonie Baken, MSc,* Melek Rousian, MD, PhD,† Erwin J. O. Kompanje, MD, PhD,‡ §§ Anton H. J. Koning, PhD,|| Peter J. van der Spek, PhD,¶ Eric A. P. Steegers, MD, PhD,# and Niek Exalto, MD, PhD**

*PhD Student and †Resident, Division of Obstetrics and Prenatal Medicine, Department of Obstetrics and Gynaecology, and ‡Senior Researcher, Department of Intensive Care, Erasmus MC, University Medical Center Rotterdam; and §§Curator, Collection Human and Non-Human Dysmorphology, Natural History Museum Rotterdam; and ||Associate Professor and ¶Professor, Department of Bioinformatics, Erasmus MC, University Medical Center Rotterdam; and #Professor and **Associate Professor, Division of Obstetrics and Prenatal Medicine, Department of Obstetrics and Gynaecology, Rotterdam, the Netherlands Netherlands

Objectives: Conjoined twins are rare. High-quality imaging techniques are essential for proper first-trimester diagnosis. Technological development leads to new imaging techniques such as 3-dimensional virtual embryoscopy. The aim of this review was to explore imaging techniques used in the first-trimester diagnosis of conjoined twins and provide a systematic diagnostic table for making this diagnosis.

Design: A PubMed literature search was performed using the terms ultrasound, Doppler, MRI, and CT combined with first-trimester and conjoined twins. Three recent cases at our department are reviewed and examined additionally using 3-dimensional virtual embryoscopy.

Results: The different types of conjoined twins are summarized in a table for practical use during ultrasound examination. In evaluating conjoined twins, 2-dimensional ultrasound is the criterion standard. Three-dimensional and Doppler ultrasounds add anatomical and prognostic information. Virtual embryoscopy imaging reveals additional findings in our 3 cases not seen with routine 2-dimensional ultrasound examination.

Conclusions: Each case of conjoined twins is unique and should be evaluated with the best possible imaging techniques. Three-dimensional and Doppler ultrasound should be added to the systematic diagnostic evaluation of conjoined twins. Virtual embryoscopy imaging may contribute to earlier, more appropriate counseling and management of these pregnancies.

Target Audience: Obstetricians and gynecologists, family physicians

This research was financially supported by Erasmus Trustfonds, Erasmus MC vriendenfonds, Meindert de Hoop foundation, and Fonds NutsOhra.

The authors have disclosed that the V-Scope software has not been approved by the U.S. Food and Drug Administration.

All authors and staff in a position to control the content of this CME activity and their spouses/life partners (if any) have disclosed that they have no financial relationships with, or financial interests in, any commercial organizations pertaining to this educational activity.

Correspondence requests to: Leonie Baken, MSc, Department of Obstetrics and Gynaecology, Division of Obstetrics and Prenatal Medicine, Erasmus MC University Medical Centre Rotterdam, Room NA15.23, PO Box 2040, 3000 CA Rotterdam, the Netherlands. E-mail: l.baken@erasmusmc.nl.

Supplemental digital content is available for this article. Direct URL citations appear in the printed text, and links to the digital files are provided in the HTML text of this article on the journal’s Web site (www.obgynsurvey.com).
Learning Objectives: After completing this CME activity, physicians should be better able to differentiate between the different types of conjoined twins and their prognosis, compare the different imaging techniques used for the evaluation of conjoined twins to decide whether additional diagnostic imaging techniques are useful in an individual case, and evaluate complex cases, such as conjoined twins, using new imaging techniques.

Conjoined twins are a rare phenomenon. These twins have fascinated people since old age; the oldest known printed illustration of a pair of conjoined twins is in the 1499 book by Jacob Locher, *Carmen heroicum de partu monstrifero*. The prevalence of conjoined twins in live births is estimated to be around 1 in 250,000. There is an unexplained female predominance (3:1).

Two hypotheses exist on the origin of conjoined twins: the fission theory in which a fertilized ovum divides incompletely and the fusion theory explaining secondary fusion of 2 originally distinct monovular embryos. Spencer argues that all types of conjoined twins can only be explained by secondary fusion. The recent finding of a monochorionic diamnionic conjoined twin pregnancy may further contribute to the fusion theory. Others are in favor of the fission theory, maintained by the observation that the incidence of mirror imaging is higher in conjoined twins than in monozygotic twins. No matter what theory is correct, conjoined twinning is an infrequent and random event challenging physicians in making a proper diagnosis, which is essential for considering treatment options.

Nowadays, conjoined twins are frequently detected during first-trimester ultrasound examinations, challenging the use of sophisticated diagnostic imaging techniques.

Proper first-trimester diagnosis of conjoined twins is important for many different reasons: to avoid false-positive diagnosis; detect related ultrasound abnormalities; the possibility to offer early termination with less maternal morbidity and less psychological impact; the possibility of selective feticide, and the reduction of coexisting conjoined twins in a triplet pregnancy or even a quadruplet pregnancy; and to monitor the associated risks of a (conjoined) twin pregnancy.

In this study, we explore the first-trimester diagnosis and typing of conjoined twins. Imaging techniques used in the diagnostic process are described. We provide a systematic diagnostic table that might be helpful in the first-trimester diagnosis of a conjoined twin.

METHODS

Literature Search

Articles were identified through a PubMed database search retrieving articles on the first-trimester diagnosis of conjoined twins. The literature search was performed in January 2013 for all available articles written in English. The free-text search terms “ultrasound,” “Doppler,” “MRI,” and “CT” in combination with “first trimester” and “conjoined twins” were used. Articles on parasitic conjoined twins were excluded. References of all relevant articles were hand-searched for additional citations.

Diagnostic Process of Recent Cases

In the past years, 3 cases of conjoined twins were diagnosed in the Erasmus MC University Medical Center Rotterdam in which informed consent for research was given by the parents. All 3 cases were initially examined using 2-dimensional (2D) and 3-dimensional (3D) ultrasound. For consecutive analysis using virtual embryoscopy (VE), the transvaginal 3D images made on the GE Voluson 730 expert and E8 system (GE Medical Systems) were converted to cartesian volumes, using specialized 3D software (4D View; GE Medical Systems) and were transferred to the BARCO I-Space Virtual Reality system. The volumes were resized, turned, and clipped in different planes to obtain the best possible images for evaluation. The evaluation of the 3 cases with Virtual Reality was performed before termination of the pregnancy and evaluated by a different examiner who was blinded to the results of the previously performed 2D/3D ultrasound.

The diagnostic findings during 2D and 3D ultrasound were extrapolated from the medical records. All findings seen with the VE imaging technique were documented and compared with the 2D/3D ultrasound findings.

RESULTS

Classification of Conjoined Twins

Each set of conjoined twins is unique with respect to the site and extent of union and their complex anatomy. The most complex anatomy is situated at the site of union. Description of conjoined twins is made easier with a classification into 8 types, advocated by Spencer. These 8 types are named cephalopagus, thoracopagus, omphalopagus, ischiopagus, parapagus, craniopagus, rachipagus, and pyopagus. The suffix “pagus” means fixed.
TABLE 1
Types of Conjoined Twins and Their Characteristics

<table>
<thead>
<tr>
<th></th>
<th>Ventral</th>
<th></th>
<th></th>
<th>Dorsal</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Cephalopagus</td>
<td>Thoracopagus</td>
<td>Omphalopagus</td>
<td>Ischiopagus</td>
<td>Parapagus</td>
<td>Craniopagus</td>
</tr>
<tr>
<td>Incidence</td>
<td>11%</td>
<td>19%</td>
<td>18%</td>
<td>11%</td>
<td>28%</td>
<td>5%</td>
</tr>
<tr>
<td>Extent of union</td>
<td>Head to umbilicus</td>
<td>Thorax to umbilicus</td>
<td>Umbilicus</td>
<td>Lower abdomen and pelvis</td>
<td>Lower abdomen and pelvis</td>
<td>Cranium (never foramen magnum/skull base)</td>
</tr>
<tr>
<td>Varieties</td>
<td>Symmetrical/asymmetrical</td>
<td>Fusion from sternum to umbilicus</td>
<td>End-to-end fusion/twins facing each other</td>
<td>Diophasus/diprosopus</td>
<td>Orientation in any position</td>
<td>Fusion into occiput</td>
</tr>
<tr>
<td>Head/face</td>
<td>Fused, 2 faces on opposite sides of head</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>1 or 2 heads, 2 faces</td>
<td>Fused skull, 2 faces</td>
</tr>
<tr>
<td>Central nervous system</td>
<td>2</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>Anencephaly</td>
<td>Fused meninges separate brains</td>
</tr>
<tr>
<td>Vertebral column</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Shoulders</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>2-3</td>
<td>4</td>
</tr>
<tr>
<td>Upper limbs</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>2-4</td>
<td>4</td>
</tr>
<tr>
<td>Thorax</td>
<td>Fused</td>
<td>Fused</td>
<td>2</td>
<td>Fused</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Heart</td>
<td>2</td>
<td>1 Conjoined/2 fused</td>
<td>2</td>
<td>2</td>
<td>1 or 2</td>
<td>2</td>
</tr>
<tr>
<td>Abdomen</td>
<td>1</td>
<td>Fused</td>
<td>Fused</td>
<td>2</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Umbilicus</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Liver</td>
<td>2</td>
<td>Shared</td>
<td>Shared</td>
<td>Minimal fusion</td>
<td>Shared</td>
<td>2</td>
</tr>
<tr>
<td>Gastrointestinal (GI tract)</td>
<td>1 Upper GI</td>
<td>2</td>
<td>Shared</td>
<td>1 Lower GI</td>
<td>1 Lower GI</td>
<td>2</td>
</tr>
<tr>
<td>Genitalia</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>Shared</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Pelvis</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>Fused</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Lower limbs</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>3-4</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>Viability</td>
<td>No</td>
<td>Rare</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Separability</td>
<td>No</td>
<td>Not likely</td>
<td>Yes</td>
<td>Yes</td>
<td>Not likely</td>
<td>Not likely</td>
</tr>
</tbody>
</table>

Bold font indicates presence in 100% of cases.
The first differentiation in this classification of conjoined twins is made in ventral versus dorsal union. Further differentiation is made by the site of union. The 8 types of conjoined twins with their characteristics, incidence, and vitality are summarized in Table 1 (Supplemental material 1, http://links.lww.com/OBGYSURV/A12).

The prognosis always depends on the specific type as described in Table 1. Surgical separation is very likely to fail with the loss of both children when a single heart is seen during first-trimester ultrasound.24

Clues to the Diagnosis

Suspicion of conjoined twinning should be raised in a twin pregnancy with a single placenta, when it is not possible to demonstrate a separating amniotic membrane or when there is only 1 yolk sac present. Signs of possible conjoined twins are also the following: more than 3 vessels in the umbilical cord, no change in relative positions of twins after movement or follow-up scans, fewer limbs than would be expected, hyperflexion of the spine, and bifid appearance of the fetal pole.2,26–28 Also increased nuchal translucency thickness can be seen in conjoined twins, especially in thoracopagus due to hemodynamic disturbances.29 When fetal activity increases, at around 8 weeks’ gestational age (GA), it becomes easier to differentiate between monoamniotic twins and conjoined twins.30

Imaging Techniques

Ultrasound

Ultrasoundography is widely used in the obstetrical field and is the most important and primary imaging technique in prenatal diagnosis of conjoined twins.2 The first diagnosis of conjoined twins using transabdominal ultrasound was in 1976.31 The first diagnosis of conjoined twins in the first trimester, at 12 weeks of GA, was reported by Schmidt et al32 in 1981.

Most diagnoses of conjoined twins are established using 2D ultrasound, especially when fusion of body parts is obvious. Ultrasound is the preferred investigation modality because it is nonionizing and noninvasive and has low costs and broad availability. It also permits real-time examination, useful as conjoined twins do not switch their relative position. Minimally conjoined omphalopagus twins can be an exception to this rule; changes in relative position have been reported.33,34

The introduction of the transvaginal ultrasonography provided ultrasound images with high resolution and therefore made it possible to visualize the early pregnancy.35 This has led to advances in first-trimester diagnosis of abnormalities in the fetal anatomy,36 like in evaluating the extent of fusion in conjoined twins.26 The earliest reported diagnosis of conjoined twins was performed using transvaginal ultrasound at 7 weeks of gestation.37

3D Ultrasound

Three-dimensional ultrasound became available with the advances made in computer technology and has shown to be helpful in the detection of congenital abnormalities.38,39 In conjoined twins, 3D ultrasound is used to exactly define the extent of fusion and to obtain more precise anatomic information.40–42 In many cases of conjoined twins, 3D ultrasound helped to confirm the presence of anomalies and improved diagnostic confidence.41–48 Especially facial features, like in parapagus or cephalopagus conjoined twins, can be studied in detail better using 3D ultrasound.43,49 Three-dimensional surface rendered images may also help the future parents to understand the complex anomalies in their fetuses.41,42,48

Doppler Ultrasound

Doppler ultrasound has expanded its application in obstetrics including in the evaluation of conjoined twin pregnancies.16,42,50 The prognosis of conjoined twins largely depends on the conjunction of the cardiovascular system. Especially the differentiation between thoracopagus and omphalopagus, to correctly classify and determine the prognosis, can be facilitated by Doppler ultrasound of the heart.37,45,51,52 Doppler ultrasound is an excellent tool in the evaluation of the vasculature of conjoined vital organs, like the liver, to determine prognosis and separability.14,50,53

Furthermore, Doppler ultrasound may reveal a characteristic “double-layer” umbilical arterial velocity waveform due to 2 separate arterial supplies in a single umbilical cord.54,55 Such a distinctive Doppler ultrasound pattern can be used as an extra diagnostic sign in conjoined twins as is proposed by Woo et al.54

Computed Tomography Scan

Only 2 publications, published in 1984 and 1990, respectively, were found on the use of computed tomography for antenatal diagnosis of conjoined twins, both in the third trimester of pregnancy.56,57

In case of conjoined twins, computed tomography can be used postpartum as a diagnostic tool to evaluate separability.58

Magnetic Resonance Imaging

Fetal magnetic resonance imaging (MRI) has already proven to be complementary to conventional obstetrical ultrasound. Especially in case of complex
anatomical anomalies, an MRI may provide additional information, like in evaluating conjoined twin pregnancies.\textsuperscript{59–62}

The literature does not provide cases of conjoined twins diagnosed in the first trimester with MRI. The earliest prenatally diagnosed conjoined twins using MRI was at 16 weeks of gestation.\textsuperscript{60}

When conjoined twins are presented late in pregnancy, MRI overcomes the limitations of ultrasound, like in the case of obesity and because of the decreasing amount of amniotic fluid. Magnetic resonance imaging provides overall assessment of the pregnancy during all 3 trimesters.

**Virtual Embryoscopy**

Virtual embryoscopy is a new imaging technique that enhances the 3D US modality using all 3 dimensions, in contrast to traditional 3D reconstructions viewed on a 2D screen. The Department of Bioinformatics of the Erasmus MC operates a fully immersive virtual reality system: the BARCO I-Space. It allows the viewers to perceive depth in and interact with 3D volumes in an intuitive manner. The V-Scope volume-rendering application creates a "hologram" of the 3D ultrasound volume to enable a so-called VE. Stereoscopic imaging allows discerning of fine details and understanding of complex relationships in the 3D volumes. We refer to previous articles for a detailed explanation of the BARCO I-Space and V-Scope.\textsuperscript{63} The innovative VE technique has already been successfully applied in prenatal medicine.\textsuperscript{64–72} Because of the improved depth perception and 3D interaction, the I-Space enables better assessment of embryonic and fetal structures.

As seen in our cases below, VE provides additional diagnostic information in evaluating complex anatomical structures in conjoined twins.

**Cases**

**Case 1**

A 30-year-old gravida 4 para 2 was referred to the Erasmus MC University Medical Center Rotterdam at 10 + 6 weeks' GA because of conjoined twins detected during routine ultrasound. Two-dimensional and 3D transabdominal and transvaginal ultrasound examination revealed a thoracopagus with fused thorax and abdomen, sharing 1 heart and liver (Table 2; Figs. 1A, B). The conjoined twins appeared to have 2 separate heads, shoulder girdles, pelvises, and 4 lower extremities. In both twins, fetal hydrops was noticed. Only 3 upper extremities could be visualized using ultrasound. Fetal karyotype revealed a normal male genotype. The parents were informed about the prognosis and decided to terminate the pregnancy. Pregnancy was terminated at 11 + 5 weeks’

### Table 2

<table>
<thead>
<tr>
<th>Case</th>
<th>1</th>
<th>2</th>
<th>3</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Type</strong></td>
<td>Thoracopagus</td>
<td>Cephalopagus</td>
<td>Parapagus</td>
</tr>
<tr>
<td><strong>GA</strong></td>
<td>10 w 6 d</td>
<td>13 w 1 d</td>
<td>11 w 6 d</td>
</tr>
<tr>
<td><strong>Head</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2D/3D</td>
<td>VE</td>
<td>Reference</td>
<td>2D/3D</td>
</tr>
<tr>
<td>Frontal bossing</td>
<td>2</td>
<td>2</td>
<td>No</td>
</tr>
<tr>
<td>Face</td>
<td>2</td>
<td>2</td>
<td>1/2</td>
</tr>
<tr>
<td>Vertebral column</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Shoulders</td>
<td>4</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Upper limbs</td>
<td>4</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Hands</td>
<td>Ulnar deviation</td>
<td>1/2</td>
<td>1</td>
</tr>
<tr>
<td>Thorax</td>
<td>Fused</td>
<td>Fused, hydrothorax</td>
<td>Fused, hydrothorax</td>
</tr>
<tr>
<td>Heart</td>
<td>1/2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Abdomen</td>
<td>Fused</td>
<td>Fused</td>
<td>Defect</td>
</tr>
<tr>
<td>Umbilicus</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Omphalocele</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Stomach</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Liver</td>
<td>Shared</td>
<td>2</td>
<td>?</td>
</tr>
<tr>
<td>Pelvis</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Lower limbs</td>
<td>4</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>General</td>
<td>Hydrops</td>
<td>Hydrops</td>
<td>Hydrops</td>
</tr>
<tr>
<td>Chromosomal</td>
<td>46, XX</td>
<td>46, XX</td>
<td>46, XY</td>
</tr>
<tr>
<td>Viability</td>
<td>Rare</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Separability</td>
<td>Not likely</td>
<td>No</td>
<td>Yes</td>
</tr>
</tbody>
</table>

*Bold font indicates case seen only with VE.*
GA using misoprostol. Intact conjoined twins were born, and macroscopic examination confirmed the diagnosis thoracopagus (Fig. 1D). The parents refused autopsy.

Using the I-Space VE system, the thoracopagus with fetal hydrops was confirmed (Fig. 1C). In addition, 4 arms and 4 legs could be distinguished easily as well as severe scoliosis in both fetuses, an abdominal wall defect, and an abnormal ulnar deviation of one of the hands.

Case 2

A 28-year-old gravida 2 para 1 was referred to our center at 13 + 1 weeks’ GA because of conjoined twins diagnosed during routine 2D ultrasound. Two-dimensional and 3D ultrasound examination revealed a cephalopagus with fusion of the heads, thorax, and part of the abdomens (Table 2; Figs. 2A, B). Two heartbeats, 1 stomach, 4 arms and 4 legs, and 1 shared umbilical cord were visualized. The fetal karyogram showed a normal female genotype.

The parents were informed about the lethal prognosis and decided to terminate the pregnancy at 14 weeks’ GA by induction. Intact conjoined twins were born, and the diagnosis of cephalopagus was confirmed by macroscopic examination (Fig. 2D). The parents did not give permission for an autopsy.

A cephalopagus can either be symmetrical (2 identical faces on opposite sides of the head) or asymmetrical (1 ‘‘normal’’ face and 1 reduced face). The back of the head, however, could not be visualized with 2D and 3D ultrasound.

Intuitive orientation in the data set with 3D VE allowed for detailed evaluation of anatomical structures (Fig. 1C). Three-dimensional VE visualized only 1 face and also 4 shoulders and an omphalocele, making the definitive diagnosis of asymmetrical cephalopagus conjoined twins.

Case 3

A 22-year-old gravida 3 para 2 presented at 11 + 6 weeks’ GA to our department after detection of conjoined twins during routine ultrasound. Conjoined twins with fetal hydrops were revealed with 2D and
3D ultrasound (Figs. 3A, B). Two separate heads and 2 separate spines were seen with fusion at the level of the thorax and the abdomen, sharing 1 heart, liver, and stomach. Two arms and 2 legs could be visualized as well as a small omphalocele. The fetuses were diagnosed as a parapagus dizephalus (Table 2). Karyotyping by chorionic villus sampling revealed a normal male genotype.

After counseling, the parents decided to terminate the pregnancy at 14 + 4 weeks’ GA using misoprostol. Autopsy findings correlated with the diagnosis parapagus dizephalus (Fig. 3E). There were 3 lungs, 1 heart, 1 set of kidneys, a shared gastrointestinal tract, and 2 equally normal-size brains present. The internal genitalia were male.

During 3D VE examination, 2 faces were seen with marked frontal bossing. There were 3 shoulders with 2 arms (Figs. 3C, D).

**DISCUSSION**

Accurate prenatal imaging is crucial in diagnosing the rare cases of conjoined twins. Even though with the classification, each conjoined twins is unique with respect to the site and extent of union and their complex anatomy. Prognosis, especially vitality and separability, should therefore be evaluated individually for each case with the best possible imaging techniques.

Ultrasound is the main imaging modality used in diagnosing conjoined twins and will remain so in the foreseeable future, because of its favorable characteristics such as accessibility and cost and the fact that in many cases the diagnosis can be made using conventional ultrasound, especially when the conjunction between the twins is obvious.

Three-dimensional ultrasound should always be included in the first-trimester evaluation of conjoined twins. Three-dimensional ultrasound can easily be performed and does not consume much extra time during the ultrasound examination. Because the spatial relationships in conjoined twins are complex and understanding them is essential for a proper diagnosis, 3D ultrasound will be of help in diagnosing these twins. Moreover, 3D ultrasound can provide extra anatomical information in several cases of conjoined twins,
especially regarding facial features. The surface rendered images of 3D ultrasound as well facilitated counseling of the parents.

Although 2D and 3D ultrasounds are sufficient in most cases of conjoined twins, they are not always successful in providing all detailed information of the complex anatomy in conjoined twins. As these details may modify the viability and separability in selected cases, other imaging techniques should also be considered. Moreover, ensuring the lethal prognosis is essential for parents in deciding to ask for a termination of pregnancy and facilitates the mourning process.

Doppler ultrasound imaging is important for the determination of viability and separability by visualizing vascular communications and in determining the number of hearts. When the vascular anatomy cannot be clearly visualized with ultrasound, the clinician should resort to Doppler ultrasound.

The lack of publications on MRI in first-trimester conjoined twin pregnancies may indicate that there is no benefit of this imaging technique in these particular pregnancies. However, MRI has emerged to be a complementary imaging technique in conjoined twins after the first trimester. The superiority of MRI as compared with ultrasound in the second and third trimesters might also apply to the first 12 weeks of pregnancy, especially because imaging techniques are in constant development. The excellent resolution of tissue composition perceived with MRI makes it a potential useful complementary tool in first-trimester diagnosis of conjoined twins. A thorough evaluation of this possible diagnostic effect could change our view on the diagnostic process of congenital abnormalities such as conjoined twins. Furthermore, the VE technique can also be applied to MRI to provide even more information.

In the 3 cases of conjoined twins referred to our clinic, we demonstrated that 3D VE provides additional and more precise anatomic information. A combination of 2D and 3D ultrasound examination and 3D VE improves the detailed morphological description and diagnosis.

FIG. 3. Case 3. Parapagus dicephalus. Two-dimensional US image showing hydrops and 2 separate heads. Three-dimensional US image showing fusion of thorax and abdomen and a single umbilical cord. I-Space volume showing the fusion of thorax and abdomen. Virtual embryoscopy also showed the presence of only 1 pelvis. Furthermore, frontal bossing and a small omphalocele were visualized. I-Space volume showing hydrops. Two vertebral columns are visualized. Ex vivo photograph confirming that only 2 upper and 2 lower extremities were present.
We conclude that conventional 2D ultrasound in most cases is sufficient in the first-trimester diagnosis of conjoined twins. The diagnostic process can be easily expanded with 3D and Doppler ultrasound to gain extra and more precise information in an efficient way. Three-dimensional VE provides additional diagnostic information in evaluating complex anatomical structures, especially when depth perception is needed, as in case of conjoined twins. This may contribute to earlier, more appropriate counseling and management of these pregnancies. As this is the first article on VE in the diagnosis of conjoined twins, more research is needed to evaluate the implementation of VE in the diagnostic process of conjoined twins and congenital malformations in general.

ACKNOWLEDGMENTS

The authors thank the Department of Pathology of the Erasmus MC University Medical Center for supplying them with the report and pictures of the autopsy and Hilly Versprille for making pictures of one of the cases. They also thank Frits Wilbrink for his contribution to the figures used in Table 1.

REFERENCES

1. Locher J. Carmen Heroicum de Partu Monstrifero. Ingolstadt: Johann Kachelofen; 1499.


