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Virtopsy in conjoined ischiopagus twins

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Summary

Purpose of investigation: To propose a multidisciplinary protocol for postmortem disclosure of complex fetal malformations, comparing ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), and autopsy in a case of conjoined ischiopagus twins. *Materials and Methods:* A screening second-trimester ultrasound diagnosed ischiopagus twins at 20 gestational weeks in a 31-year-old woman without any previous ultrasound examination. The couple decided for pregnancy termination. The formalin-fixed fetuses underwent full-body CT, MRI, and autopsy. *Results:* ultrasound accurately diagnosed ischiopagus twins. CT was very accurate in the description of bone components. MRI allowed better visualization of the visceral organs than CT. Only autopsy could disclose the aspect of the two gastrointestinal tracts and the external genitalia. *Conclusions:* Prenatal ultrasound represents the standard diagnostic exam for conjoined twins. CT-MRI virtual autopsy (virtopsy) may be an option if the couple refuses to authorize necropsy or may be useful to plan a minimally invasive autopsy preserving the external phenotype.

Key words: Autopsy; Computed tomography; Conjoined twins; Magnetic resonance imaging; Ultrasound; Virtopsy.

Introduction

Conjoined twins are rare and in particular, ischiopagus represents only 1.8% of them [1, 2]. According to the site of junction, there are several classifications. Nevertheless, each case of conjoined twins is unique because of the presence of anatomical variants and malformations not related to the site of union. Etiology and embryology are uncertain. The obstetrician's aim is to obtain an early and accurate diagnosis, offering to the couple a more appropriate counseling about prognosis and feasibility of postnatal separation.

Prenatal ultrasound represents the standard diagnostic exam. In addition, high-quality imaging techniques, such as three-dimensional and Doppler ultrasounds, may contribute to earlier and more accurate first-trimester diagnosis [3-5]. Especially in later pregnancy, magnetic resonance imaging (MRI) may be useful to provide a better topographical description of the malformation through its higher contrast resolution and its larger field of view. Ultrafast imaging sequences, such as the single-shot fast spin-echo T2-weighted sequence, can acquire high-quality images, minimizing motion artifacts without fetal or maternal sedation [6, 7]. Actually, the clinician should use the best possible imaging techniques to obtain an early and proper diagnosis, to better support the couple in making important decisions.

Planning of postnatal surgical separation can rely on several imaging modalities depending on the site of junction. Nevertheless, computed tomography (CT) often represents the most accurate examination in preoperative work-up. Contrast studies may be useful to disclose the anatomy of distal intestinal and genitourinary tracts [8, 9].

In cases of fetal demise, postmortem CT and MRI are able to realize a sort of virtual autopsy, so-called "virtopsy". It may represent an option to disclose fetal anatomy if the couple refuses to authorize the necropsy or may be useful in autopsy planning with specimen preservation [10-12].

Reporting the present case of conjoined ischiopagus twins, the authors would like to propose a multidisciplinary protocol for postmortem disclosure of complex fetal malformations, such as conjoined twinning. Their diagnostic protocol integrates prenatal ultrasound imaging with CT-MRI virtopsy and autopsy findings.

Case Report

A 31-year-old Caucasian woman (gravida 2, para 0) with a previous first-trimester miscarriage and without any history of consanguinity or toxic exposure underwent a screening second-trimester ultrasound, that diagnosed conjoined ischiopagus twins at 20 gestational weeks. No previous ultrasound examination was performed. The twins were fused starting from the insertion of a single umbilical cord (Figures 1a and 1c). There were four kidneys in the usual position connected with only one common bladder (Figures 1b and 1d). It was impossible to identify the external genitalia. The other body districts appeared to be normal and separated.

The couple, after counseling, refused prenatal MRI and decided for pregnancy termination. The abortion was obtained by labor induction and vaginal delivery in a breech presentation at 21 weeks of gestation. There were no maternal complications.

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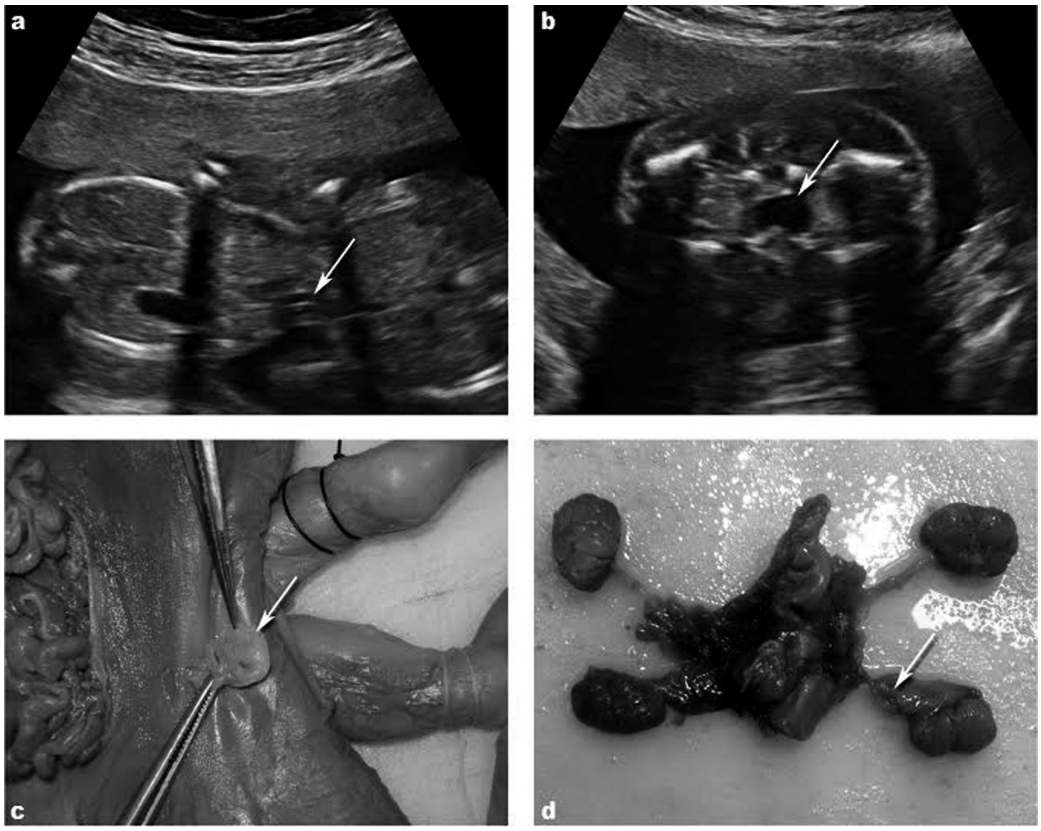

Figure 1. — Ultrasound and pathological findings. (a) Transabdominal ultrasound scan at the level of insertion of the single umbilical cord (white arrow). (b) Ultrasound scan demonstrating the single common bladder (white arrow). (c) Gross appearance of the insertion of the single umbilical cord (white arrow) with four vessels: two arteries and two veins. (d) Dissected pelvis with the four kidneys converging into a single bladder with an incomplete septum. One of the four ureters was dilated with signs of megaureter (white arrow).

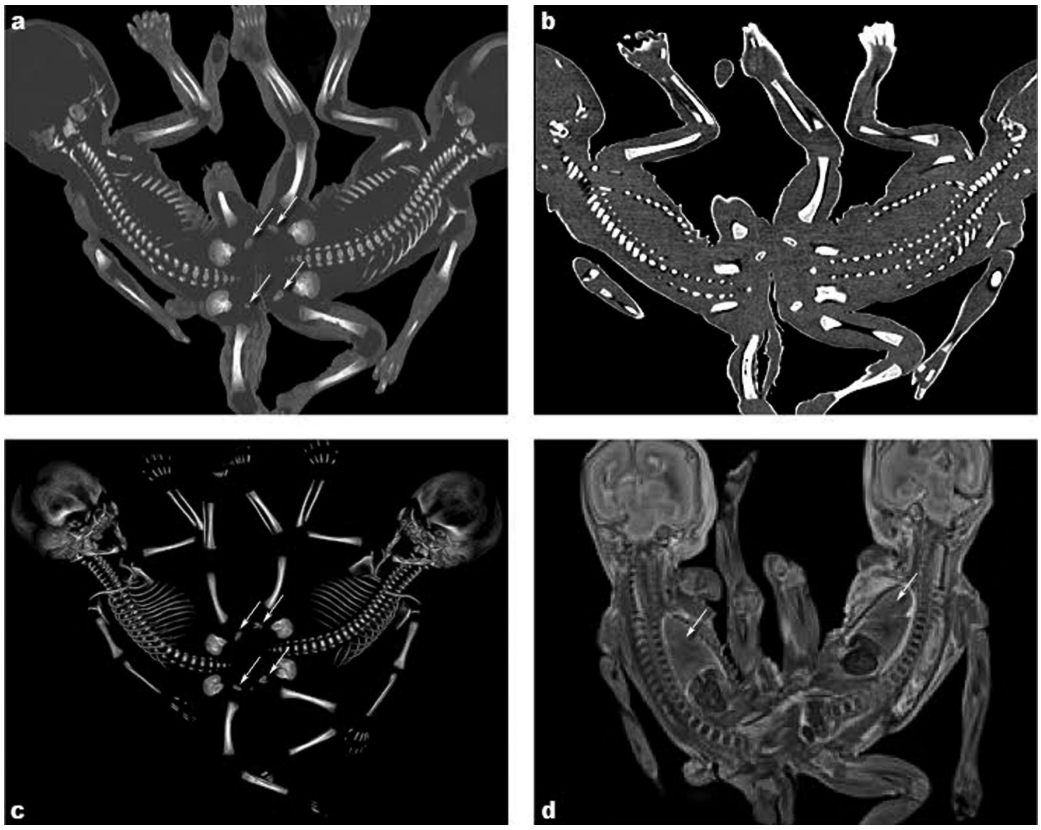

Figure 2. — Radiological findings. (a) CT scan with bone window setting demonstrating the presence of all the ossification nuclei of pelvis and femur heads in both twins (white arrows). (b) CT multi planar reconstruction showing the absence of any junction site at bone level. (c) Shaded surface-display (SSD) reconstruction of the fetal skeleton confirmed the presence of all the pelvic ossification nuclei of each fetus (white arrows). (d) T2 MRI image visualizing fetal visceral organs, as fetal lungs (white arrows).

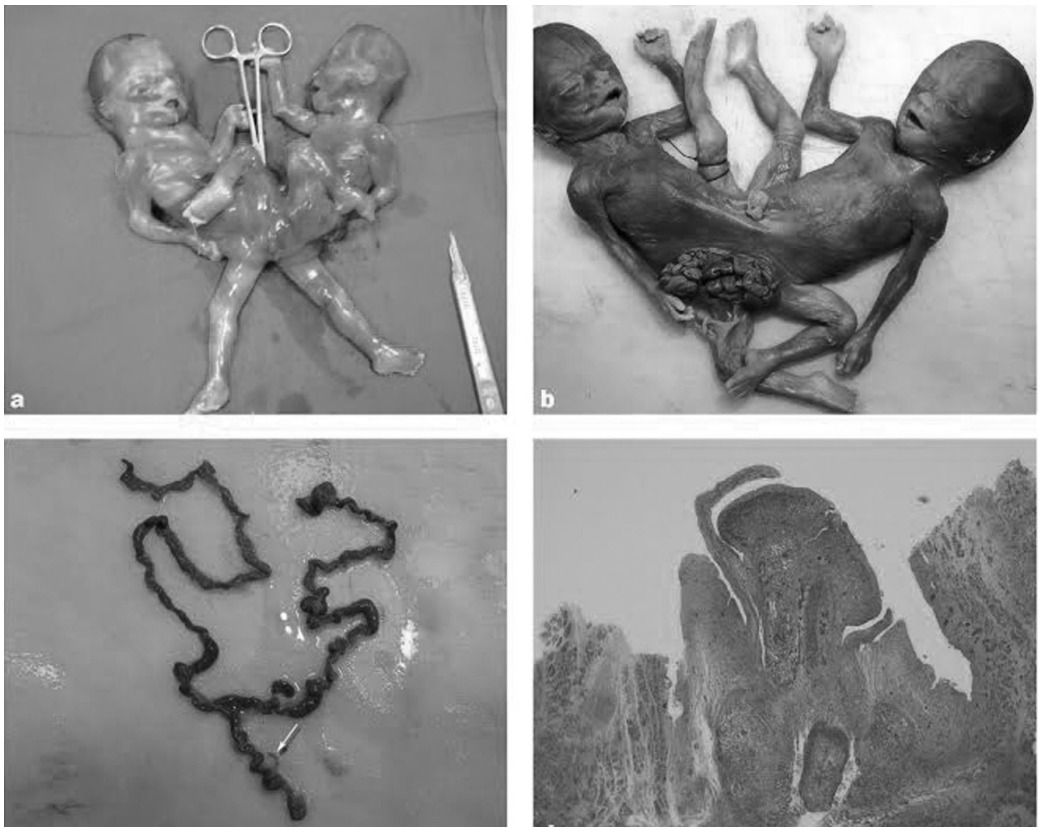

Figure 3. — Pathological findings. (a) Gross appearance of the twins just after the delivery showing signs of maceration. (b) Macroscopic features of the formalin-fixed twins with iatrogenic disruption of the anterior abdominal wall and protrusion of bowel loops. (c) The two dissected bowels with a common terminal ileum and a single large intestine with one cecal appendix (white arrow). (d) External genitalia of the conjoined twins with hypoplastic penis (Hematoxilin/ Eosin ×20).

The formalin-fixed fetuses underwent full-body CT and MRI. CT scans were acquired using a 128-slice multidetector scanner. The images and post-processing were carried out through volumetric acquisition with scans of about one mm in thickness and subsequent three-dimensional, maximum intensity projection, and multiplanar reconstructions on Syngo.via and TeraRecon 3.6.2.3 Acquarius workstations. CT was very accurate in the description of bone components, demonstrating the absence of any junction site at bone level (Figures 2a, 2b, and 2c).

MRI was performed using a high-field scanner with a superficial coil. Both bi-dimensional and three-dimensional T2-weighted sequences were performed. MRI allowed better visualization of the visceral organs with respect to CT, due to the higher tissue and contrast resolution (Figure 2d).

The conjoined fetuses weighed 614 grams. There was a single placenta of 570 grams and a single umbilical cord of 40 cm with two arteries and two veins (Figure 1c). Autopsy of the formalin-fixed twins confirmed the ultrasound and radiological findings (Figures 3a and 3b). In addition the gastrointestinal tracts were joined in a "y" seven cm above the ileocecal valve (Figure 3c). Only microscopic examination disclosed two joined prostates and a hypoplastic penis with a hypospadic urethra (Figure 3d).

Discussion

In the clinical evaluation of the present case of conjoined ischiopagus twins, the authors used a multidisciplinary approach similar to that applied to a case of gnatho-thoracopagus twins by Salmaso *et al.* [12].

In the present authors' experience, a first-trimester diagnosis was not possible, because the woman performed her first ultrasound screening at 20 gestational weeks. However, the second-trimester ultrasound was able to properly diagnose the type of conjoined twins and to exclude any other malformations not related to the site of union.

After counseling, the couple refused prenatal MRI and decided for abortion. The woman was nulliparous and desired to avoid hysterotomy. Hence, a late-second-trimester termination of pregnancy was successfully performed through labor induction and vaginal delivery, as reported by Mitchell *et al.* [13].

Virtopsy confirmed prenatal ultrasound findings, adding interesting information about skeletal anatomy. Postmortem CT and MRI were useful to provide a more accurate topographical description of fetal anatomy, disclosing the spatial relationships between anatomical anomalies and normal structures. In particular, CT disclosed fetal bone anatomy excluding any junction site at this level. Nevertheless, only the autopsy could clarify the appearance of the two bowels and the aspect of the external genitalia.

In the present authors' experience, virtopsy was useful in autopsy planning and in particular provided important information to disclose fetal bone anatomy. Moreover, the diffusion of the virtopsy protocol may improve the use of high resolution imaging techniques, such as prenatal MRI

or CT, for assessing the feasibility of postnatal separation in preoperative diagnosis [14, 15].

In conclusion, each case of conjoined twins is unique and a maximum effort should be made to obtain an early and proper diagnosis, providing the couple a more appropriate counseling. In this way, the diagnostic approach should be multidisciplinary, using the best possible imaging techniques to disclose these complex malformations. In cases of fetal demise, CT-MRI virtopsy may be an alternative method to necropsy or may be useful to plan a minimally invasive autopsy preserving the external phenotype.

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