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Imaging of liver fusion and vascular intra-hepatic shunts in omphalopagus conjoined twins: The role of a double CT study

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ABSTRACT

Omphalopagus twins are joined ventrally in the umbilical region. In omphalopagus twins, liver's fusion is very frequent, being present in about 80% of cases. Two conjoined twins born in our Hospital were evaluated using plain x-rays, ultrasound (US), gastrointestinal xrays, Tc-99 hepatobiliary scintigraphy (SC) and contrast enhanced computed tomography (CT). There was no bony connection on conventional xrays, neither bowel communication on GI x-rays study. US demonstrated the fusion of left hepatic lobes and a not-quantifiable vascular shunt.

Normal biliary function was demonstrated by Tc-99 SC. The first CT scan (twin A) showed irregular lobules in the site of hepatic fusion, a portoportal shunt and a venous vessel to the inferior vena cava of twin B. Only the second CT scan (twin B) showed an arterio-arterial partiallyextrahepatic shunts. The double CT scan allows to increase the probability of a correct individuation and description of vascular shunts for an effective preoperatory assessment. The omphalopagus twins in our case underwent planned surgical separation at 5 months of age. The surgery lasted 5 h and was not affected by any intraoperative complications.

1. Introduction

Conjoined twins are, by definition, monozygotic, monoamniotic and monochorionic. The estimated prevalence in the literature varies widely, from 1: 50,000 to 1: 200,000 [1–4].

Omphalopagus twins are joined ventrally in the umbilical region; the lower thorax may be included with the possibility of a shared pericardium, but the heart is never involved [5]. The stomach and proximal small intestine are usually separate; however, in 33% of cases, the small intestine joins at the level of the Meckel diverticulum in the distal ileum [2].

In omphalopagus twins, liver's fusion is very frequent, being present in about 80% of cases [2]: a correct preoperative assessment must provide a careful identification and description of vascular shunts among the twins.

Various imaging modalities have been shown to be useful to evaluate the area of shared parenchyma such as plane x-ray, biliary scintigraphy, ultrasonography or gastrointestinal (GI) x-ray; among these, CT has been recognized as the imaging modality of choice for an accurate description of vascular communications potentially present in omphalopagus twins [1–6].

However, the paucity of the cases does not allow to assess the best technique of execution of the CT scan, and in the literature is reported the possibility of performing either a single study or two separate ones [6].

We report a case of omphalopagus conjoined twins in where the site, typology and entity of complex vascular shunts were evaluated in the context of the shared portion of hepatic parenchyma, using a double CT study preceded by an evaluation by a different imaging method.

Since conjoined twins have unique hemodynamic features, the presence of vascular shunts with a pressure gradient must be carefully considered. Consequently, we decided to perform a double CT scan to increase the chance of a correct identification and description of vascular shunts, which are needed for an effective preoperatory assessment.

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Fig. 1. Postnatal plane x-ray performed immediately after delivery.
The x-ray demonstrates the absence of bone connections despite the close proximity of the not-yet-ossified xyphoidal processes.

2. Case report

A prenatal diagnosis of conjoined twins was accomplished through a prenatal ultrasound performed at a gestational age of 12 weeks and confirmed as omphalopagus by a fetal MRI at 14 weeks of gestation.

The twins, with a total birth weight of 4700 g, were delivered at 34 weeks +4 days of gestation by caesarean section, both with an Apgar scores at 1 and 5 min of 6 and 9.

Due to the absence of the natural compressive effect on the thoracic structures during the passage of the fetus through the birth canal, a first evaluation of the pulmonary parenchyma was performed at birth with a plain x-ray, showing no early signs of aspiration of amniotic fluid or bone segments shared between the twins (Fig. 1).

An abdominal ultrasound study was performed two days after birth (Logiq E9, General Electric, Boston, Massachusetts, US). The exam, despite the difficulties due to the mutual position of the twins, confirmed the presence of a liver parenchymal bridge (Fig. 2a), the presence of two distinct hepatic hila with two venous portal axes recognizable, separate and with hepatopetal normal flow.

The hypothesis of the presence of intraparenchymal vascular communication was set during the ultrasound study and was confirmed after the administration of ultrasound contrast medium (0.5 ml, Sonovue 8 μl/ml, Bracco Diagnostics, Milano, Italy) to the twin A. The contrast-enhanced ultrasound (CEUS) showed a venous shunt within the shared liver parenchymal bridge, which was characterized by an inhomogeneous contrastographic effect and irregular lobulatures (Fig. 2b).

A gallbladder was present in twin A, but not in twin B. There was no dilatation of the biliary tracts. Two normal kidneys were present in both twins, as well as the spleen and the pancreas.

Gastrointestinal (GI) x-rays exams performed in separate occasions did not show shared GI tracts among the twins (Fig. 3a–b).

CT scans were acquired at the age of 4 months with a multislice CT (Brilliance 64, Philips, Eindhoven, Netherlands) with a slice thickness of 2.5 mm, reconstruction thickness of 0.67 mm, pitch of 0.891, rotation time of 0.5 s, Field of View (FOV) of 180 mm; tube voltage 100 kV; with automated tube current modulation. We performed two CT examination overall, injecting one twin at the time.

The twin A was administered with 12 ml of non-ionic contrast material (Iomeron 350 mgI/ml; Bracco Diagnostics, Milan, Italy) at the dosage of 2 ml/kg, intravenously through a peripheral venous line with injection rate of 0.5 ml/s. We acquired unenhanced, arterial and portal venous phases, including both thorax and abdomen. The total dose of these scans (DLP) was 451.7 mGy$*$cm^2.

Three days later we injected the twin B with 10 ml of the same non-ionic contrast material (Iomeron 350 mgI/ml; Bracco Diagnostics, Milan, Italy) at the dosage of 2 ml/kg, intravenously through a peripheral venous line with injection rate of 0.5 ml/s. We performed the arterial and the portal venous phase, on thorax and abdomen, with a total dose (DLP) of 277.6 mGy$*$cm^2.

Overall, the total dose (DLP) to the twins was of 729.3 mGy$*$cm^2 (CTDI vol. 28.46 mGy).

The diagnostic capability of the CT study was reinforced by multiplanar, MIP and 3D elaborations with the use of different image management protocols.

The twins showed atrial situs solitus, without evidence of visceral heterotaxia on CT.

The CT scan non-EKG gated, showed an interatrial defect in twin A (Fig. 4), confirmed by cardiac ultrasound.

The first CT scan (twin A) showed irregular lobules in the site of hepatic fusion, a porto-portal shunt and a venous vessel to the inferior vena cava of twin B (Fig. 5a–b). Only the second CT scan (twin B) showed an arterio-arterial partially-extrahepatic shunt, probably related to a pressure gradient between the twins (Fig. 6a–b). A separate hepatic venous drainage into the inferior vena cava and right atrium, which is critical for survival after separation surgery, was clearly demonstrated on both CT.

The liver volume and the degree of parenchymal enhancement of twin B appeared smaller than that of twin A (Fig. 7). As previously showed by ultrasound, the gallbladder was normally identified on CT in twin A and not clearly identified in twin B; therefore a TC-99 hepatobiliary scintigraphy was performed.

The examination was performed during a dynamic acquisition for 60 min from the administration of the radionuclide(99mTc-Mebrofenina) and subsequently with SPECT/CT acquisitions of the abdomen (Siemens, Symbia T).

The activity/time curves relating to the cardiac, hepatic, cholecystic and upper intestinal areas were reconstructed.

Scintigraphy demonstrated regular hepatobiliary function and established the presence of two gallbladders (Fig. 8).

3. Discussion

Thanks to advances in imaging, anesthetic and surgical techniques, prognosis of omphalopagus twins is one of the most favorable among conjoined twins.

Preoperative assessment should be aimed at demonstration of anatomical fusion, vascular anomalies including vascular shunts and cross-circulation, and associated abnormalities that are important for surgical planning, which in turn largely determine the successful surgical separation and post-surgical survival.

These essential preoperative imaging findings can be accurately obtained by performing a double CT study.

Abdominal MRI in omphalopagus twins is limited by long examination time and the respiratory motion artifacts, larger in newborns

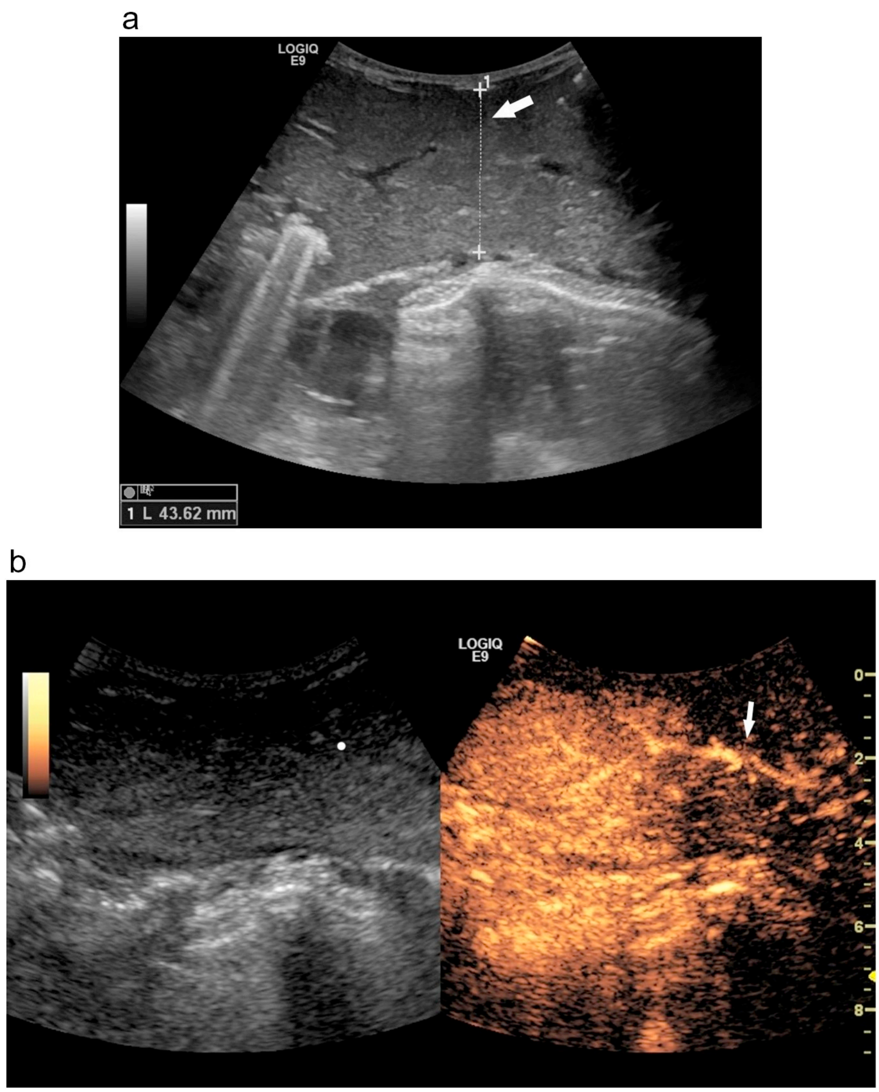

Fig. 2. Ultrasound study (a) of the abdomen performed two days after birth (white arrow) shows the parenchymal liver bridge with a sub-xyphoidal extension. Ultrasound performed with contrast medium (b) questions the presence of venous shunts within the shared parenchymal bridge (white arrow), at the level of which there is a inhomogeneous contrastographic effect.

because of their proportions. Moreover, the position of the twins creates a challenge for imaging on MRI – a lower spatial resolution modality compared with CT. Also, the choice of coils would be difficult for postnatal imaging in these conditions.

CT has been suggested as the most promising non-invasive imaging modality for evaluating omphalopagus twins [1,4–6].

In our case, abdominal CT was successfully performed in sedated omphalopagus infants, without the need of endotracheal intubation or general anesthesia.

Some authors recommend for contrast-enhanced CT two separate intravenous injections of contrast agent into each twin on different days [6]; this of course will double the CT radiation dose, but we chose this approach since we wanted to precisely describe the altered and common vascular anatomy of the twins and the resulting hemodynamic characteristics.

The clinical evaluation of the patients did not highlight morphological parameters suggestive for the presence of a continuous unidirectional shunt between the twins (similar to a twin-twin transfusion).

The possibility of an arterio-arterial communications made it necessary to perform a double angiographic CT study to limit the influence of any pressure gradient between the twins.

Furthermore, delayed-phase CT scan provided useful information regarding anatomy and function of parenchymal organs such as the liver, spleen and kidneys [1].

Contrary to what has been described in the case of thoracophagus twins [7], a classification of the degree of involvement of the hepatic parenchyma and of the corresponding vascular shunts in the omphalopagus twins has not yet been proposed, although it may constitute a valid element to be correlated with the surgical outcome.

In our opinion, in the case of omphalopagus conjoined twins any assessment should consider at least 5 elements:

- the site of the vascular connections within the shared parenchyma for a correct guide to the surgical incision;
- the typology of the vascular shunts (porto-portals, arterio-arterial, porto-systemic) for a prediction of intraoperative risk of bleeding;

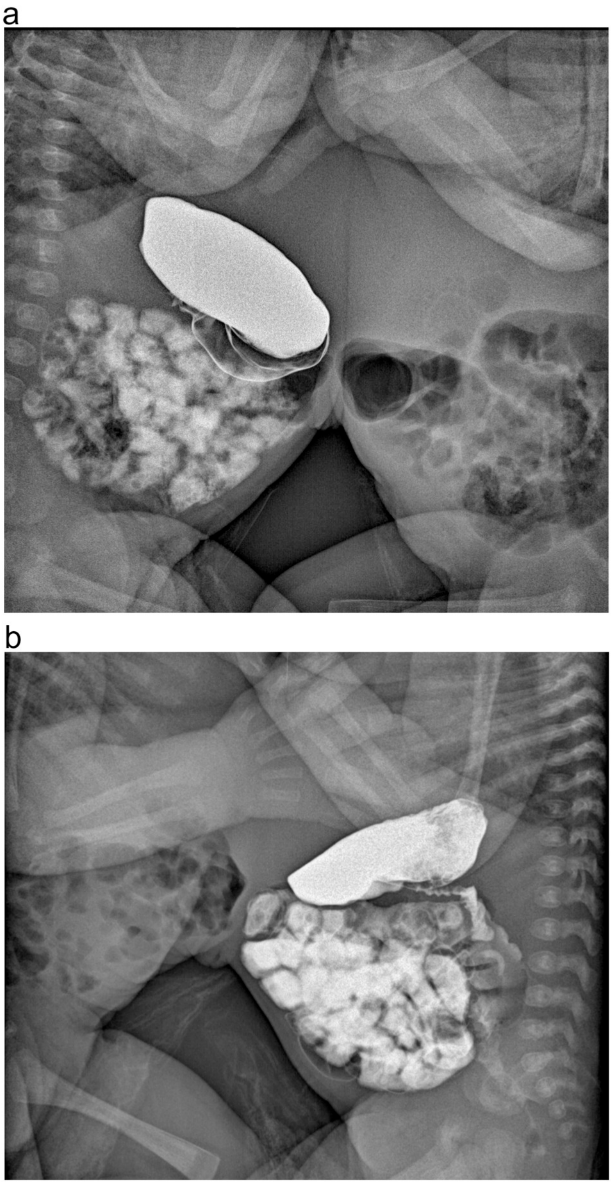

Fig. 3. a–b. Gastrointestinal x-ray studies confirm the presence of an independent GI system per twin with no shared tracts among the twins.

Fig. 5. Axial view (a) of the CT scan performed on twin A showed irregular lobules (*) in the site of hepatic fusion and a venous vessel to the inferior vena cava of twin B (white arrow).
Sagittal view (b) shows a porto-portal shunt (white arrowhead) within the shared hepatic parenchyma.

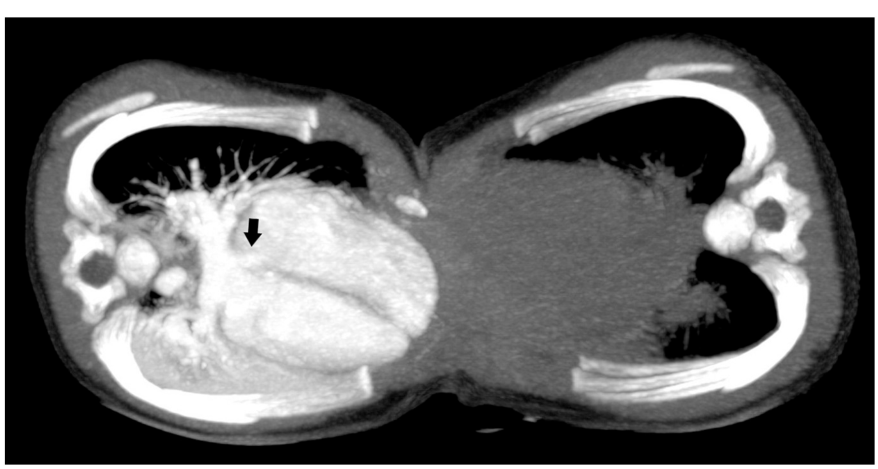

Fig. 4. Atrial situs solitus with a not-before detected interatrial defect (black arrow) and without evidence of visceral heterotaxia on CT.

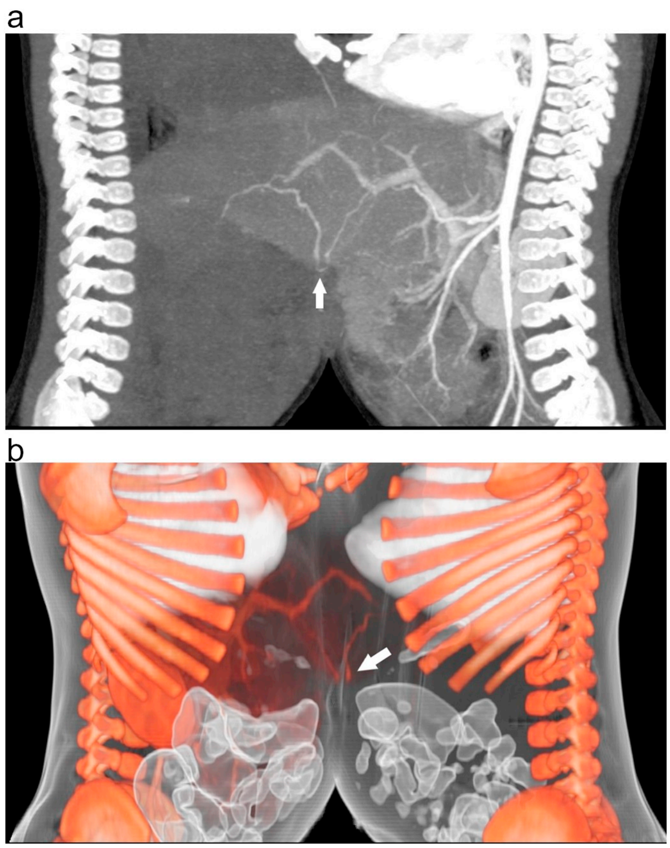

Fig. 6. CT scan performed on twin B: MIP reconstruction (a) and 3D volume rendering (b) show the partially-extrahepatic arterio-arterial shunt (white arrow), probably related to a pressure gradient between the twins.

- the parenchymal volume remaining in each twin after separation for a correct prediction of liver function;
- the arterial and venous vascularization of the shared parenchyma portion to avoid the postoperative risk of the onset of ischemia;
- the presence or absence of normal liver function tests.

Additionally, a complete imaging and physical examinations should be performed to guarantee the best chance of success of the subsequent twin separation intervention.

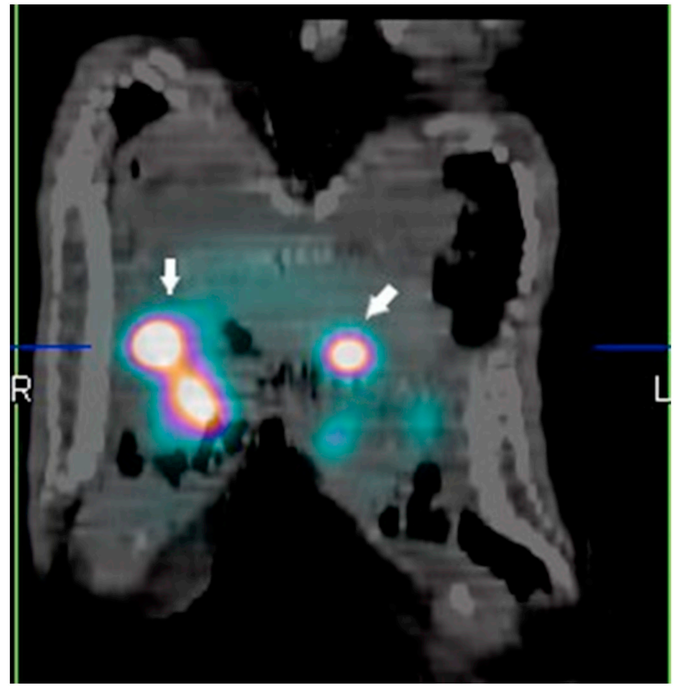

Fig. 8. The presence of two gallbladders (white arrows) is proved by TC-99 hepatobiliary scintigraphy performed for 60 min from the administration of the radionuclide (99mTC-Mebrofenina) and subsequently with SPECT/CT acquisitions of the abdomen.

Therapeutic options include conservative nonsurgical management, emergency separation during the neonatal period, and planned separation, usually at 6–12 months of age.

The omphalopagus twins in our case underwent planned surgical separation at 5 months of age to promote the correct development of the axial skeleton. The surgery (Fig. 9a–b) lasted 5 h and did not lead to any intraoperative complication.

The post-operative period was characterized by the clinical stability of the twins, until their discharge 6 months and 15 days after delivery.

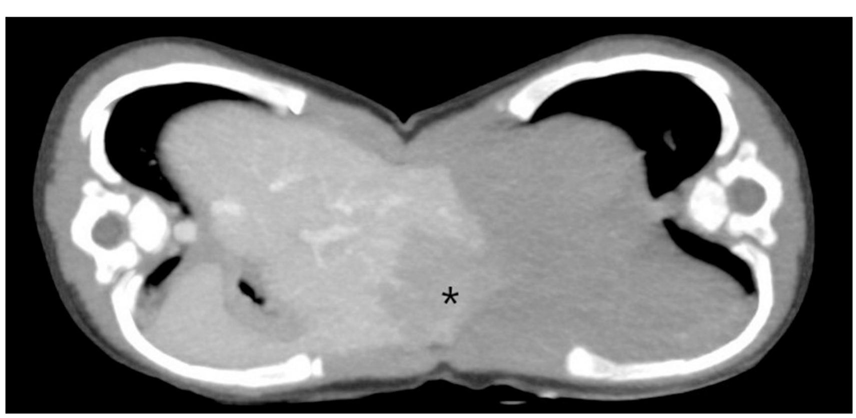

Fig. 7. CT scan performed on twin B (on the left) showed a smaller liver volume as compared to the twin A (on the right) and the degree of parenchymal contrast enhancement of twin B (*).

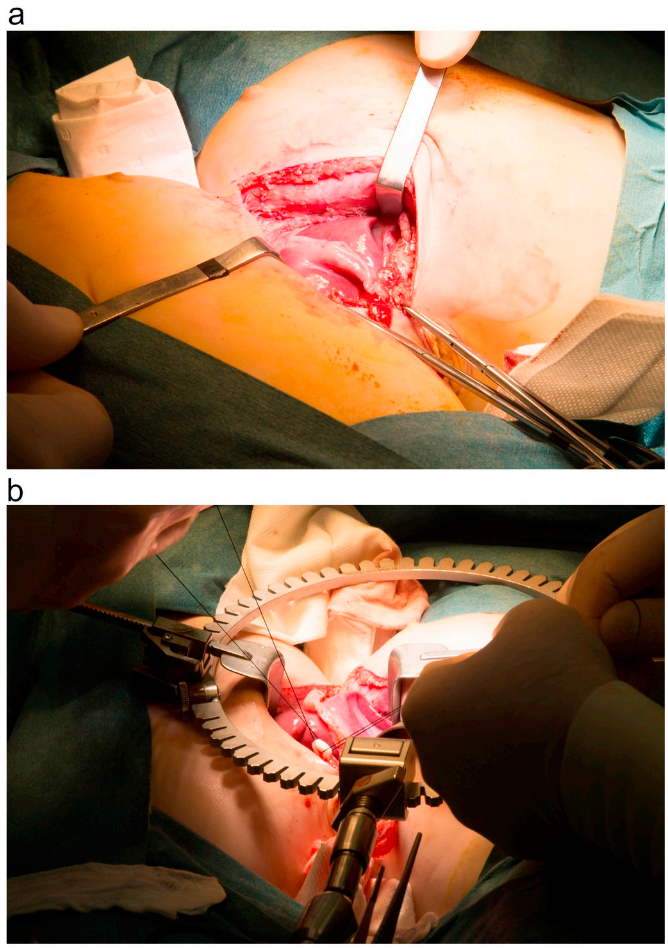

Fig. 9. Intraoperative findings of hepatic parenchymal bridge (a) and the surgical evidence of the venous vascular shunt between the twins (b).

Sources of support

None.

Declarations of interest

None.

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