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Separation of pygopagus, omphalopagus, and ischiopagus with the aid of three-dimensional models

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ABSTRACT

Background: The three-dimensional (3D) technique provides with accurate anatomical information. We present the separation surgeries for three different kinds of conjoined twins with the aid of three-dimensional techniques.
Method: For the pygopagus twins, a pelvic and lower vertebral model was made. For the omphalopagus and ischiopagus, their enhanced computed tomography (CT) scan images were transferred to the Computer-Assisted Surgery Planning System (CASP) (Hisense, Qingdao, China) to generate the 3D models.
Result: In the case of the pygopagus twins, the 3D model clearly showed that their coccyges were joined at a 120°angle from each other horizontally which suggested that the blind-end orifice on their back was a pilonidal sinus, which separated the normal sphincter into two halves. In the omphalopagus, the 3D model revealed one of the branches of each twin's hepatic vein was connected with the other's. The 3D model of the ischiopagus twins revealed that both of the twins had duplicated bladders and each baby's duplicated bladders united with one of the bladders of the other baby and a single rectum passing through the middle of the four bladders.
Conclusion: 3D techniques could provide more detailed anatomical information, which is helpful in planning procedures for such complicated separation surgery.
Levels of evidence: Level IV.

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Conjoined twins are the result of a rare congenital anomaly, developing from incomplete duplication of a single fertilized ovum in the process of monozygotic twinning. The incidence of conjoined twins is estimated to be about 1 in 50,000 births [1,2]. The widely-used classification of conjoined twins is according to the prominent site of fusion, which was proposed by Spencer [3]. Due to the variable and complex anatomic structure of the fused part, detailed imaging examination is of great importance for surgical planning. Three-dimensional (3D) modeling technology has been used to assist with various aspects of surgery, especially in delineating the anatomic structure and drawing up the surgical plan. We report here that successful separation of conjoined twins of different kinds – pygopagus, omphalopagus and ischiopagus – with the assistance of three-dimensional modeling from February 2015 to February 2016. Informed consents for these separation surgeries were obtained. The Ethics Committee of the Children's Hospital of Fudan University approved these separation surgeries.

1. Patients and methods

1.1. Pygopagus

Female pygopagus twins were delivered by cesarean section at 38 weeks. They were transferred to our neonatal intensive care unit at 3 days after birth. Initial evaluation revealed healthy-appearing conjoined twins joined at the sacrum, buttocks and perineum (Fig. 1A). Each of them had a separate urethra and vagina. There were two orifices arranged vertically on either side of the vaginas, one of which defecated while the other orifice was a blind end (Fig. 1B). Barium contrast imaging of the GI tract showed they had two GI tracts that shared a 0.5 cm common anal canal (Fig. 2A). Magnetic resonance imaging (MRI) revealed that their sacra and coccyges were joined and their dural sacs seemed continuous, but magnetic resonance imaging urography revealed normal anatomy. The separation surgery was scheduled for 3 months later, and during this time the parents were trained to dilate the common anal canal.

When the twins were readmitted, we performed a further examination of their perineum under anesthesia. With electrical stimulation, it was revealed that each of the two orifices (the defecating one and the blind-ended one) had a semi-circle of muscle around the orifice which

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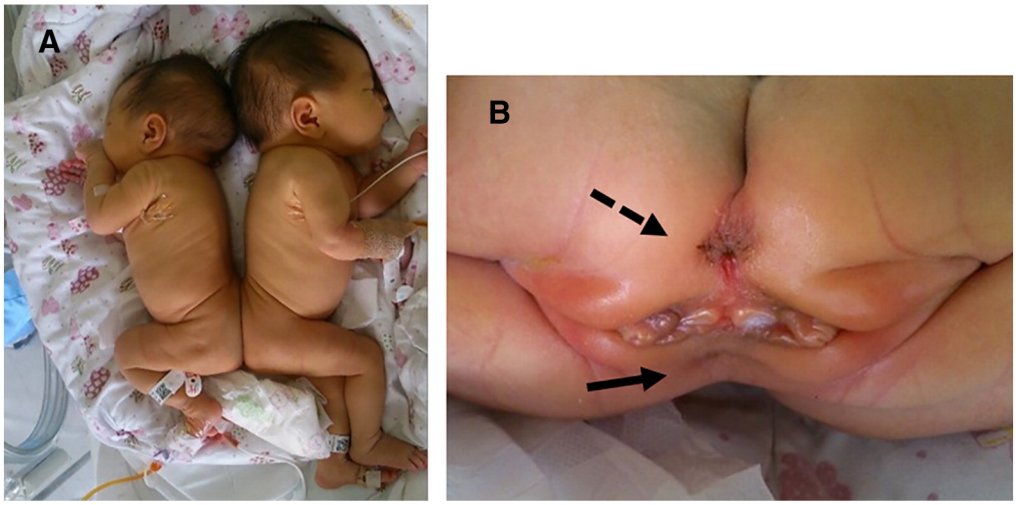

Fig. 1. (A) Pygopagus conjoined twins joined at the sacrum, buttocks and perineum. (B) The orifice that defecated is shown by the dashed arrow; the blind-end orifice is shown by the solid arrow.

could contract normally. We expected that these two semi-circles of muscle that contracted normally could be used to reconstruct two anuses, but only one would have normal function and the other one would be incontinent. Computed tomography (CT) was performed to delineate the anatomy of the joined part, which showed there were no great vessels communicating within the shared perineum. Based on the CT images, a bony pelvic and lower vertebral plastic model was made (Fig. 2B), which clearly showed that their sacra were separated but the coccyges were joined at a 120° angle from each other horizontally. This 120° angle caused the blind-end orifice to be positioned on their back and near the coccyx, which suggested that the blind-end orifice was a pilonidal sinus that separated the normal sphincter into two halves. Thus, we expected that we could incise the blind-end orifice in the middle, and use these muscles and those surrounding the orifice for defecation to construct two anuses. In this way, we could maximally restore the two babies' anal function. A plastic model of the abdomen, lower back, buttocks and pelvic skin was made in order to design the flap.

1.2. Omphalopagus

Male omphalopagus symmetrical twins (Fig. 3), weighing 4150 g, were delivered by emergency cesarean section at 34 weeks + 5 days. They were transferred to the neonatal intensive care unit of our hospital because of anhelation. Physical examination showed that they were joined from the xiphoid to the umbilicus. A Doppler study showed that their hearts were separate with no life-threatening malformations. Abdominal MRI revealed that they shared a liver, but there were two portal triads. After receiving one month's treatment, their condition became stable and they were discharged. Separation surgery was planned for 3 months later.

In order to delineate the anatomy of the liver and associated vessels by 3D modeling, contrast-enhanced abdominal CT with arterial-phase, portal-phase and delayed-phase was performed for each baby separately. All the three-phase images were transferred to the Computer-Assisted Surgery Planning system (CASP) (Hisense, Qingdao, China). The CASP segmented the anatomical structure of the liver, postcava and intrahepatic vessels and then generated the liver 3D model. The 3D model revealed that their livers were fused and that there was an interpenetrating plane in the middle of their fused liver. In addition, one of the branches of the hepatic vein of each twin connected with the other's, which was confirmed by CT images showing that one baby's hepatic vein was visualized in the other baby's venous phase. Further, the model revealed that some hepatic veins passed through the fused plane of their livers which would need to be ligatured in the separation surgery. More precisely, it showed that when Twin A was on the left, we would ligature the right branches of the portal vein of Twin A in the fused place at first, and then their connected middle hepatic veins, and the left branches of Twin B′s portal vein followed (Fig. 4). In addition, a plastic model of the twins' joined abdomens was created to help the plastic surgeon to calculate the skin they shared and design the flap.

1.3. Ischiopagus

Male ischiopagus conjoined twins (Fig. 5A), weighing 5140 g, were delivered by cesarean section at 36 weeks + 6 days. When they were

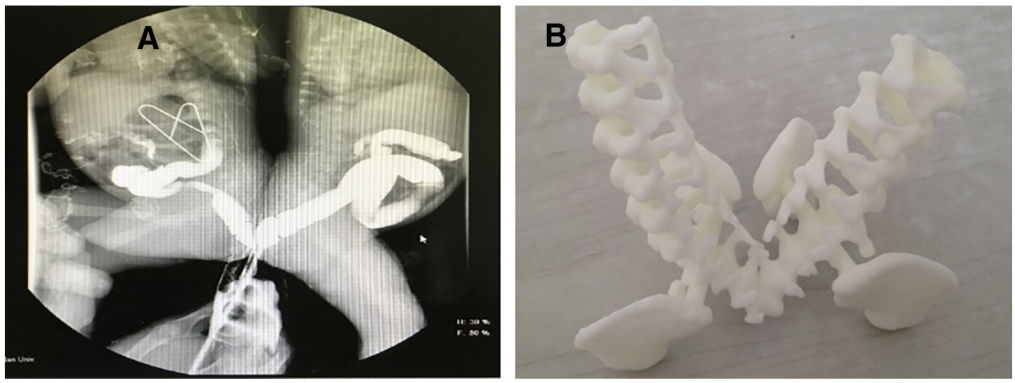

Fig. 2. (A) A barium contrast study showed they had two GI tracts that shared a 0.5 cm common anal canal. (B) The plastic model of the bony pelvis and vertebrae, which clearly showed that their coccyges were joined at a 120° angle from each other horizontally.

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