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CASE REPORT

Thoraco-omphalopagus asymmetric conjoined twins: Report of a case and complete review of the literature

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Summary Thoraco-omphalopagus asymmetric conjoined twinning is a rare condition in which a grossly defective foetus (the parasite) is attached to the thorax and upper abdomen of the main foetus (the autosite). We describe a case of thoraco-omphalopagus asymmetric conjoined twins in which the autosite had an associated large-diameter omphalocoele that was successfully separated at our institution. Reconstruction of the resulting abdominal-wall defect was performed using a flap from the gluteal region and the proximal portion of the inferior limb of the parasite, which is demonstrated. In addition, a review of all previously published cases is presented, showing that overall positive results can be obtained in treating this condition and that the presence and degree of cardiac involvement have a major influence on the prognosis.

Asymmetric or heteropagus conjoined twins (also known as parasitic twins) are an extremely rare occurrence. Some large retrospective studies have indicated an incidence of one case out of 1—2 million births.[1,2] The condition is defined as the presence of a severely defective foetus (or foetal parts) attached to a relatively normal twin (termed the autosite) in one of the same areas in which intact conjoined twins would be united.[3]

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Thoraco-omphalopagus twins are those whose area of fusion is located between the upper thorax and the umbilicus of the autosite. A marked predominance of the male gender is observed and other malformations are frequently associated with this condition, such as cardiac abnormalities and omphalocoele.[3]

Being such a complex and rare occurrence, the surgical approach always poses a challenge for the surgical team, especially when facing the need for closure of a large defect. We present a case of successful surgical treatment of a thoraco-omphalopagus twin with a safe option for reconstructing a large abdominal defect, along with a review of all previously published cases.

This work was fully approved by the Ethics Committee of the Faculty of Medical Sciences, Campinas State University — Unicamp.

Case report

A male full-term baby born at our Obstetrics and Gynecology Hospital was the first child of a 22-year-old healthy mother. The gestational period was uneventful except for non-treated asymptomatic bacteriuria. An ultrasonography exam performed at 26 weeks had diagnosed heteropagus twinning, and because of this diagnosis a scheduled caesarean section was elected as the safest method of delivery. The neonate had a total weight of 3975 g and Apgar scores of 6 and 9.

The parasite twin was attached to the anterior thoraco-abdominal wall of the autosite. There were two well-formed upper limbs with two hands and nine fingers, connected to the upper third of the thoracic wall, at the level of the manubrium. At the level of the inferior thoracic wall and epigastrium arose the inferior abdomen and pelvis of the parasite, continued by two well-formed lower limbs, with feet and 10 toes. There was also a pervious anus, which passed meconium, and male genitalia consisting of a penis and a bifid scrotum (Figure 1). The penis contained a normally positioned urethra and there was urine flow through it. The four limbs had no active movements and nor did they react to stimuli.

As associate malformations, the autosite presented an 8-cm-large omphalocoele and a partial and deformed duplication of the right ear. Echocardiography showed a patent foramen ovale.

A computed tomography (CT) scan revealed that arterial supply to the parasite came from a wide branch arising from the brachiocephalic trunk of the autosite. The scan could also anticipate most of the intra-operative findings related to the parasite's contents and its anatomical relation to the autosite.

The baby's parents were informed in detail about the condition and the need for a surgical procedure and also that the parasite, by definition and based on the preoperative findings, was not a viable sibling of the autosite. Explanations were given on the differences between a parasite and a symmetric conjoined twin. The intervention received then the parents' consent.

After thorough evaluation and planning, surgical separation of the twins was performed by the paediatric and plastic surgery teams at the 23rd day of life. An incision was initially made around the lower abdominal segment of the parasite and continued down to the omphalocoele, entering the abdominal cavity. The parasite had a complete urinary system, with two kidneys, ureters and bladder. Two undescended testes were found in the parasite's cavity. Further, there was a rectum and a segment of colon which communicated with the autosite's ileum. Two livers were found in the autosite's abdominal cavity, each one with its own gallbladder, and were left in site. The arterial branch coming from the autosite's brachiocephalic trunk could be seen entering the parasite and giving rise to what would be its aorta and subsequently dividing into the iliac branches. Removal of the parasite's viscera, pelvic bones and left-lower limb, maintaining the above-mentioned main vessels, was performed by the paediatric surgery team. As it was felt that the abdominal defect could not be primarily closed without tension, we developed a flap using tissue from the right gluteal region and thigh of the parasite by disarticulating the extremity at the level of the knee and removing the right femur through a medial incision. From the initially obtained area of the flap, a portion was maintained based on blood perfusion, as a clear distinction between well and poorly perfused areas could be made intra-operatively (Figure 2). The abdominal defect was then closed in a tension-free manner, with the uppermost part of the thoracic incision closed primarily.

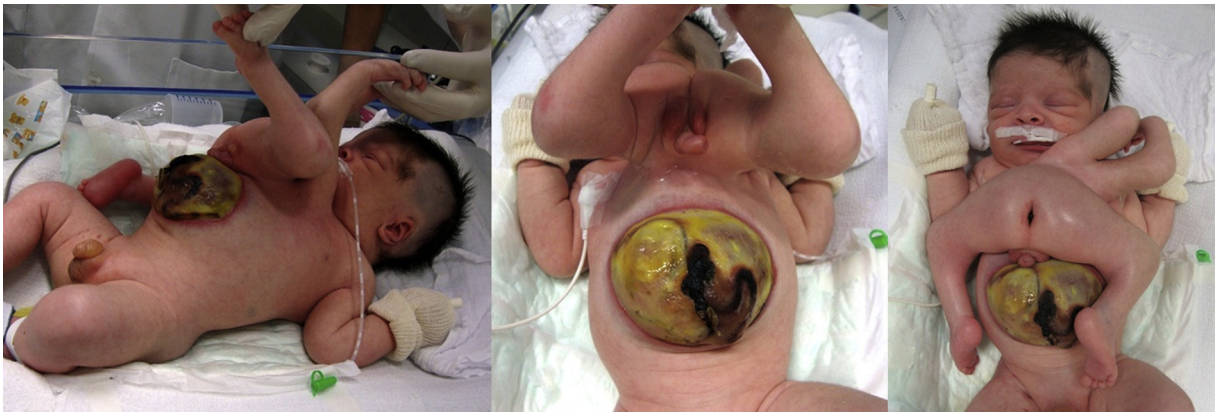

Figure 1 Preoperative aspect of the newborn with parasite attached to the thoraco-abdominal wall and a large omphalocele.