

Ischiopagus tetrapus conjoined twin with Gastroschisis

Renu Singh, Yashodhara Pradeep,
Pooja Gupta

Department of Obstetrics and
Gynaecology, Chhatrapati Shahuji
Maharaj Medical University, Lucknow,
Uttar Pradesh, India

Abstract

Parasitic conjoined twins are rare anomalies of monochorionic monoamniotic twins. We report a case of ischiopagus tetrapus conjoined twin with single head, and absent anterior abdominal wall. Due to its variable anatomy no definite treatment plan is available. Early prenatal diagnosis assists in management.

Introduction

Conjoined twins are rare with an estimated frequency of 1 in 50,000 in utero to 1 in 250,000 live births.¹ Parasitic twins are asymmetric conjoined twins in which the severely defective twin (parasite) is attached to the fully developed body of the co-twin (autosite). It is further classified into (i) an externally attached parasitic twin (ii) an enclosed foetus in fetu (iii) an internal teratoma (iv) a cardiac connected via the placenta.² It is an extremely rare condition with an estimated incidence of less than 0.1 in 100,000 births. Prenatal diagnosis of conjoined twins with B mode and three dimensional ultrasound, computed tomography (CT), magnetic resonance imaging (MRI) has been reported.³ Prenatal diagnosis of parasitic twins is relatively difficult because of the changing fetal positions and the rarity of these congenital anomalies. However, only few cases of parasitic conjoined twins have been diagnosed in utero.⁴ We report a case of conjoined twin with their spine fused in the lower part with gastroschisis, single heart with congenital heart defect, multiple limbs, and single head suspected on prenatal ultrasonography. Following termination of pregnancy, the conjoined twins were ischiopagus tetrapus with gastroschisis.

Case Report

A 26-year-old Indian woman, para 2 was referred to our tertiary care centre at 20 weeks

of gestation after ultrasonographic suspicion of conjoined twins with gastroschisis. It was a spontaneous conception. Her previous two pregnancies had been uneventful and the children were alive and healthy. There was no history of consanguinity or of twinning in the family. There was no history of maternal tobacco, alcohol or illicit drug use. Her obstetric ultrasound done for the first time at 20 weeks of gestation showed grossly deformed twin foetuses with spine fused in the lower part (Figure 1A). There was a single abdomen with deficient anterior abdominal wall suggestive of gastroschisis. Head could only be visualised in one foetus. A single heart was visualised with presence of ventricular septal defect. Multiple fetal limbs were also seen with talipo equinovarus deformity. The amniotic fluid was normal. The parents were counseled about the poor prognosis of the fetal anomaly. As the gestational age was 20 weeks, they opted for termination of pregnancy and an ischiopagus tetrapus twin with single head and gastroschisis was delivered. On examination, the co-twin (autosite) had normal facial features with developed head. The two upper limbs were on either side of the thorax. The thorax was short with herniation of the gastrointestinal organs. Two lower limbs, at right angles to the trunk were seen on the either side of the site of union, with talipes deformity. The parasitic twin was acephalic and attached to the autosite at the caudal end. Upper limbs, two in number were present on either side with deformities. Two lower limbs at right angles to trunk were also seen on the either side of the point of union. The genitalia of both were male and were laterally placed between the legs. The abdomen was a single one with herniation of its contents (Figure 1B). Single perineal opening was seen on the back (Figure 1C). The parents did not give consent for autopsy due to religious reasons and hence the internal anatomy could not be confirmed. X-ray showed a single O shaped pelvis, the two sacrum faced each other, and there was a division of the symphysis pubes (Figure 1D).

Discussion

Conjoined twinning is a rare aberration of monozygotic monoamniotic twinning. It is thought to occur either because of incomplete fission of the blastocyst inner cell mass during the primitive streak stage or fusion of two originally distinct inner cell masses that coalesce at a later stage. In parasitic twinning, vascular compromise has been proposed as the cause of development of asymmetry between the autosite and parasite. Factors like maternal age, ethnicity, maternal drug, alcohol use or smoking are not associated with the develop-

Correspondence: Renu Singh, Type IV/11, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Raebareilly road, Lucknow, Pin 226014, Uttar Pradesh, India.
Tel: 091.05222495188.
E-mail: narayanrenu@yahoo.com

Key words: twins, ischiopagus, tetrapus, gastroschisis.

Acknowledgements: the authors would like to thank Dr. Manju Tandon, for referring this interesting case to their centre.

Contributions: RS, manuscript drafting and preparation; YP, manuscript critical evaluation and final approval; PG, literature searching.

Conflict of interest: the authors report no conflicts of interest.

Received for publication: 16 June 2011.
Accepted for publication: 24 August 2011.

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Teratology Studies 2011; 1:e1
doi:10.4081/ts.2011.e1

ment of asymmetric conjoined twinning. Family history of multiple gestations was reported in only 3 parasitic twin cases.⁵ Multiple instances of symmetric conjoined twinning developing in association with assisted reproductive technologies^{6,7} and one case of parasitic twinning after intracytoplasmic sperm injection has been reported.⁸

Epidemiological data regarding parasitic twinning is scarce. A recent European study shows an incidence of parasitic twinning of 0.02 per 100,000 million births.⁹ Omphalopagus (union between the thorax of one twin and umbilicus of the other twin) is the most frequent variant in parasitic twinning. The ischiopagus variety is rarer in which the twins are joined at the pelvis. Ischiopagus twins are subdivided according to the number of lower extremities.¹⁰ Ischiopagus tripus is the commonest variety in which the lower extremities on one side are represented by a composite limb. The rarest is the bipus variety with no lower extremities on one side. Tetrapus is a subtype in which all four lower extremities are present and are oriented at right angles to the axis of the common trunk. Though the internal anatomy is variable, coupling of the gastrointestinal tract occurs at Meckel's diverticulum, distal small and large intestine usually become a common tract and have dual blood supply. The upper urinary tract is most commonly duplicated. They also manifest an array of

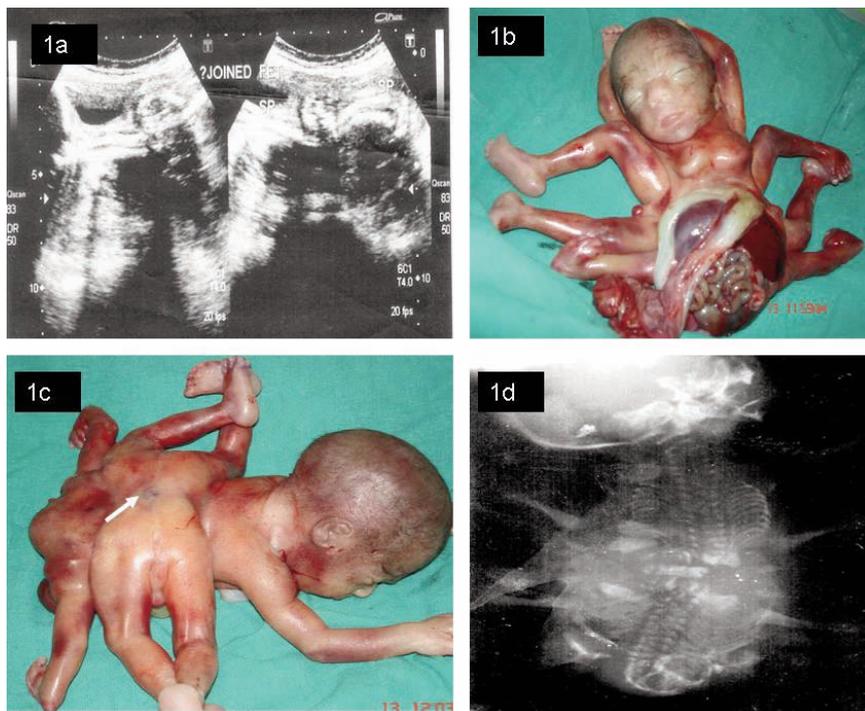


Figure 1. A) Prenatal ultrasonography showing twin foetuses with fused spine at lower end; B) Anterior photograph of the delivered fetus. The autosite (1) and the parasite (2) with single abdomen with herniation of its contents including stomach, liver, spleen and the intestines; C) Single perineal opening seen on the back of Ischiopagus tetrapus twin; D) X-ray showing a single O shaped pelvis, the two sacra facing each other, with division of the symphysis pubes.

spinal abnormalities like scoliosis and spina bifida. The normally closed circular pelvis is widely open as a semicircle resulting in the fusion of each pubic symphysis with the corresponding symphysis of the opposite twin. Thus the bony pelvis is circular but comprised of two separate, fused pubes.¹¹ In our case the parasitic twin was ischiopagus tetrapus with gastroschisis. Spencer has reported gastroschisis in only two atypical ischiopagus twins in his review of 120 ischiopagus twin cases.¹² Presence of gastroschisis in parasitic twins signifies early embryological disturbance.

Survival is generally good among cases of parasitic twins because of less extensive visceral and vascular connections between the autosite and parasite. Presence of congenital

cardiac defect is associated with poor prognosis.⁵ There are very few reports of successful separation of ischiopagus tetrapus twins, where the baby was delivered at term.^{10,11,13} In situations where the opportunity of antenatal diagnosis of this condition is missed, at birth, careful evaluation and planning are important for subsequent operative approach. Early prenatal ultrasound is essential for the diagnosis of this severe condition.

Conclusions

Ischiopagus tetrapus twins are rare forms of conjoined twins. Prenatal diagnosis assists in antenatal management.

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