

# A rare variety of ischiopagus tetrapus conjoined twins

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## SUMMARY

Ischiopagus is a very rare variety of conjoined twins with very few cases reported in the literature. A case of ischiopagus-tetrapus conjoined twins is reported here. Twins were fused at the lower halves of the bodies. There were four well-formed separate lower limbs. The twins had two anal openings for passage of both urine and stool. In general, female predominance is found in conjoined twins. But this particular case, where external genitalia were absent making gender determination difficult, is even more unusual among the rare type. Follow-up of the twins was not possible, as they passed away during pre-operative procedure.

**Key words:** Ischiopagus – Tetrapus – Conjoined twins – Genitalia agenesis

## INTRODUCTION

Conjoined twinning is one of the rare, most interesting and most challenging congenital malformations (Ghritlaharey, 2008). Conjoined twins occur with the incidence of 1 in 50,000-100,000 births, and only 6-11% of the conjoined twins are of the ischiopagus type (Sarin et al., 1998). Ischiopagus twins are joined at the pelvis and they usually have three (tripus) or four (tetrapus) limbs (Hoyle et al., 1989). About half of the ischiopagus have four separate lower limbs, 1/3rd have 3 lower limbs (2 separate and 1 fused) attached to the

body laterally and 1/5th cases are parasitic (O'Neill et al., 1988). Conjoined twinning arises when the twinning event occurs at about the primitive streak stage of development, or 13-14 days after fertilization, and is exclusively associated with the monoamniotic monochorionic type of placentation (Nyundo et al., 2013). The exact cause of conjoined twinning remains unclear. Some authorities consider it the result of incomplete division of the embryonic disc, while others opine that there is secondary fusion of two originally separate monozygotic embryonic discs (Rode et al., 2006).

A rare variety of ischiopagus tetrapus type of conjoined twins with associated agenesis of external genitalia is discussed herein.

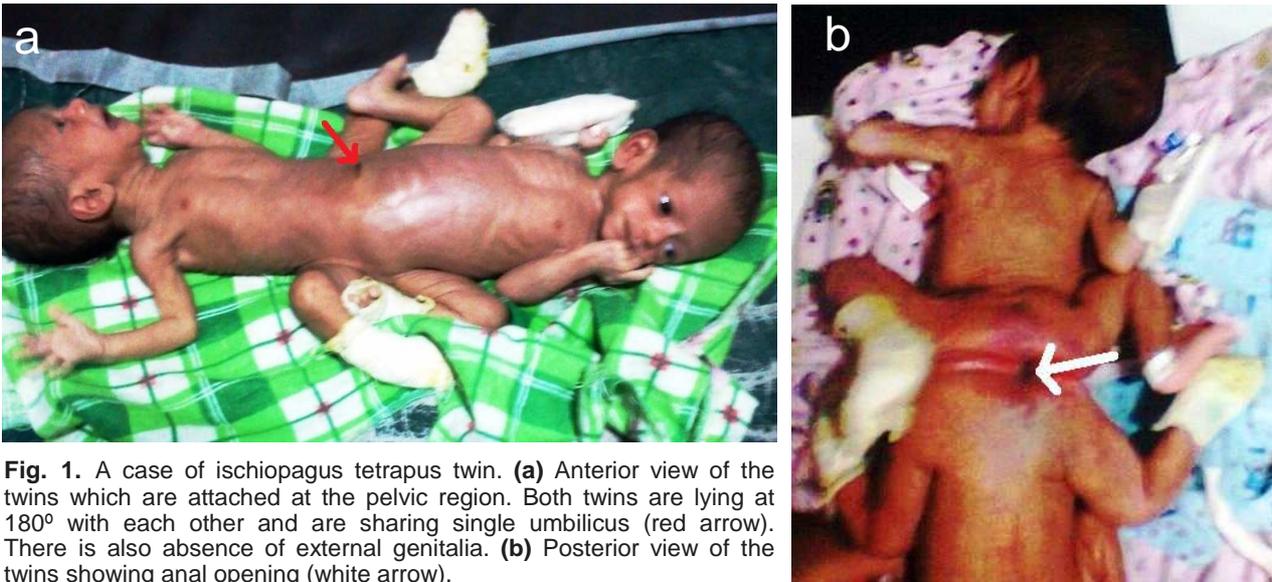
## CASE REPORT

A pair of conjoined twins fused at lower halves of the bodies with four lower limbs (Fig. 1a) was referred to the Radiology Department for pre-surgical investigation. The twins were full-term with a combined birth weight of 4.5 kg, and were delivered vaginally to a 30-year-old female who didn't undergo any antenatal ultrasonography. There was no family history of twinning, but consanguinity was present. History of radiation exposure or intake of drugs was absent. Informed consent was taken from the guardians for examination and investigations.

On external examination, both the babies were alive and apparently well developed (Fig. 1a). The two babies were joined at the pelvic and perineal regions with the vertebral axis at 180° with each other. Both twins had normal heads, necks and upper limbs. Each twin had its own pair of lower limbs oriented at right angles to the axis of the

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**Fig. 1.** A case of ischiopagus tetrapus twin. **(a)** Anterior view of the twins which are attached at the pelvic region. Both twins are lying at 180° with each other and are sharing single umbilicus (red arrow). There is also absence of external genitalia. **(b)** Posterior view of the twins showing anal opening (white arrow).

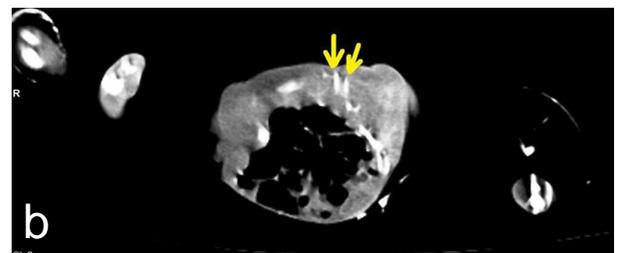
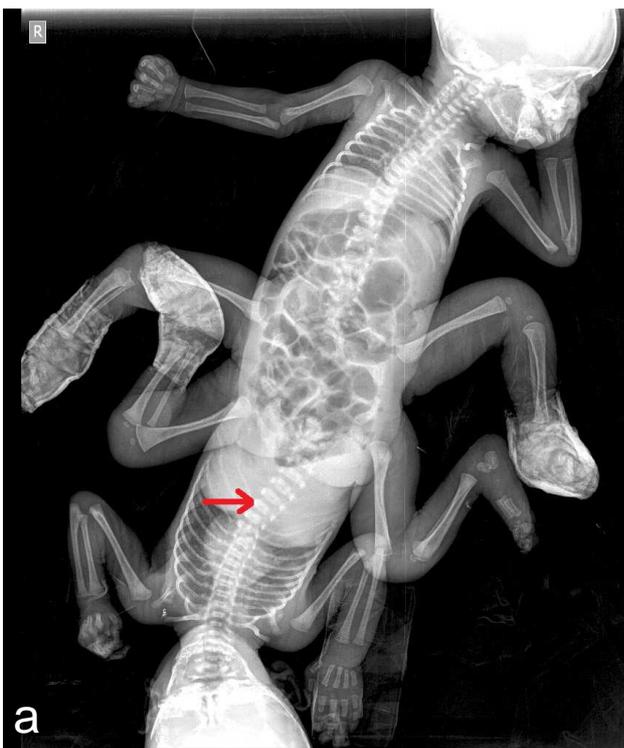
common trunk. A single umbilicus was placed ventrally in the midline (Fig. 1a). External genitalia was absent for both twins. Two anal openings were observed dorsally in the midline at the fused region of the twin for passage of both urine and stool (Figs. 1b and 2b). No separate urethral opening was present.

The X-ray revealed fusion of the pelvic region and showed two separate spines and pelvic bones. Scoliosis of dorso-lumbar spine was noted in one of the neonate (Fig. 2a). No bony fusion of lower extremities was observed. There was no evidence of any other vertebral anomalies. The contrast enhanced computed tomography (CECT)

scan, performed in a Philips 16 Slice CT scanner (with injection iomeron 350 i.e iomeprol 71.44g corresponding to 35g of iodine at concentration 350mg iodine per ml), revealed two anal openings which were placed dorsally (Fig. 2b). The sacrum, coccyx and ilium part of the two pelvises of both twins was noticed with no bony fusion between them. A complete CECT study was not possible because the twins died the very next day. The attendant refused any further postmortem dissection and investigation.

**DISCUSSION**

Conjoined twins are most challenging patients faced by pediatric surgeons because of their rarity, variable anatomy, and no definite treatment (Qazi et al., 2002). The incidence of conjoined twins is higher in Indian and African races, and is defined as 1:14,000-25,000 live-births (Ezike et al., 2010). Robertson studied conjoined twins and determined the incidence of various types as being 73% thoracopagus, 19% pygopagus, 6% ischiopagus and 2% craniopagus (Robertson, 1953). The ischi-



**Fig. 2.** **(a)** Radiograph of the ischiopagus conjoined twins showing mild scoliosis of one twin (red arrow). There is absence of bony fusion between the pelvises. **(b)** Contrast enhanced computed tomography (CECT) with dorsal surface of the twins faced upward, showing two anal openings (yellow arrows).

opagus variety of conjoined-twins is the most complicated form of twinning, with significant organ sharing (Khan, 2011). Tetrapus is a subtype of ischiopagus in which all 4 lower extremities are present and oriented at right angles to the axis of the common trunk (Zhang et al., 2007).

It is reported that one of the twins is almost always weaker or smaller, though genetically identical and may have additional congenital defects (Rode et al., 2006). Khan and Sangari also reported cases of ischiopagus twins with one neonate smaller than the other (Khan, 2011; Sangari et al., 2001). However, in the present case both the twins grossly were of equal size. About 51% of the ischiopagus twins have shared pelvic organs. Urinary bladder may be single or double, lying side by side or combined with one bladder draining into the other (Rode et al., 2006). In this case, the twins had two perineal openings which were draining both urine and stool.

The common abnormalities seen in conjoined twins are pulmonary atresia, transposition of great vessels, defect or absence of atrial or ventricular septae, many intestinal abnormalities including imperforate anus and severe scoliosis. (Golladay et al., 1982). In the present case, one of the twins showed scoliosis. Female predominance in the order of 3:1 is found among conjoined twin (Chalam, 2009). Cases of ischiopagus reported by other authors had complete or incomplete external genitalia (Pal et al., 2014; Khan, 2011; Sangari et al., 2001). But there was absence of external genitalia in the present case, thus indicating it to be a rare variety.

It is possible to diagnose conjoined fetuses by antenatal routine two-dimensional trans-vaginal ultrasound examination as early as 9<sup>th</sup> week of gestation (Basgül et al., 2006). Because of the highly variable and complex anatomy and associated malformations, skilled clinical assessment aided by detailed radiological studies, appropriate planning and team work are required for the successful separation of the conjoined twins (O'Neill et al., 1988).

## CONCLUSION

Ischiopagus is a rare variety of conjoined twins, and associated agenesis of external genitalia makes the case a rarer one. Antenatal evaluation in the form of ultrasonography and subsequent management are the preventable measures for such cases.

## REFERENCES

- BASGÜL A, KAVAK ZN, SEZEN D, BASGUL A, GOKASLAN H (2006) Thoraco-omphalopagus conjoined twins detected at as early as 9 weeks of gestation: trans-vaginal two dimensional ultrasound, color Doppler and fetoplacental Doppler velocity waveform findings. *Fetal Diag Ther*, 21: 477-480.
- CHALAM KS (2009) Anaesthetic management of conjoined twins' separation surgery. *Indian J Anaesth*, 53: 294-301.
- EZIKE HA, AJUZIEOGU VO, AMUCHEAZI AO, EKENZE SO (2010) General anesthesia for repair of omphalocele in a pair of conjoined twins in Enugu, Nigeria. *Saudi J Anaesth*, 4: 202-204.
- GHRITLAHAREY RK (2008) Ischiopagus Tetrapus conjoined twins. *J Clin Diagn Res*, 2: 800-801.
- GOLLADAY ES, WILLIAMS GD, SEIBERT JJ, DUNGAN WT, SHENEFELT R (1982) Dicephalus Dipus conjoined twins. A surgical separation and review of previously reported cases. *J Pediatr Surg*, 17: 259-263.
- HOYLE RM, THOMAS CG (1989) Twenty-three-year follow-up of separated ischiopagus tetrapus conjoint twins. *Ann Surg*, 210: 673-679.
- HAN YA (2011) Ischiopagus tripus conjoined twins. *APSP J Case Rep*, 2: 5.
- NYUNDO M, JAHN A, KAYONDO K, NTIRENGANYA F, MUZUNGU K, NTAKIYIRUTA G, KIVIRI W, KARANGWA JN, KAYINAMURA JB, BAHATI A, BYIRINGIRO JC, MUTESA L, GASHEGU J (2013) Successful surgical separation of conjoined twins: First experience in Rwanda. *East Central Afr J Surg*, 18: 124.
- O'NEILL JA, HOLCOMB GW III, SCHNAUFER L, TEMPLETON JM, BISHOP HC, ROSS AJ III (1988) Surgical experience with thirteen conjoined twins. *Ann Surg*, 208: 299-310.
- PAL R, ARORA KK (2014) Colostomy in an ischiopagus, 3rd PND conjoined twins with cross-circulation: Anaesthetic management. *Indian J Anaesth*, 58: 345-347.
- QAZI AQ, HAQ AU, BURKI T, KHAN A, MANSOOR K, CHAUDHRY A (2002) Separation of xiphi-omphalo-ischiopagus tetrapus twins with favorable internal anatomy. *J Pediatr Surg*, 37: 9.
- ROBERTSON EG (1953) Cranipagus parietalis, report of a case. *Arch Neurol Psychiatry*, 70: 189-205.
- RODE H, FIEGGEN AG, BROWN RA, CYWES S, DAVIES MRQ, HEWITSON JP (2006) Four decades of conjoined twins at Red Cross Children's Hospital - lessons learned. *S Afr Med J*, 96: 931-940.
- SANGARI SK, KHATRI K, PRADHAN SJ (2001) Omphalopagus ischiopagus tetrapus conjoined twins - a case report. *J Anat Soc India*, 50: 40-42.
- SARIN YK, MAHAJAN JK (1998) Ischiopagus tetrapus conjoined twins. *Indian Pediatr*, 35: 773-774.
- ZHANG XS, FENG ZG, XIONG QX, LI MJ, TANG DX (2007) Successful separation of ischiopagus tetrapus conjoined twins. *World J Pediatr*, 3: 228-231.

BASGÜL A, KAVAK ZN, SEZEN D, BASGUL A, GOKASLAN H (2006) Thoraco-omphalopagus conjoined twins detected at as early as 9 weeks of gestation: trans-vaginal two dimensional ultrasound, color Doppler and fetoplacental Doppler velocity waveform