Eur Spine J
DOI 10.1007/s00586-014-3707-y

CASE REPORT

Surgical management of pygopagus conjoined twins with spinal bifida

Rahul Awasthi · Raghvan Iyengar · Shrikant Rege · Nilesh Jain

Received: 2 September 2014 / Revised: 28 November 2014 / Accepted: 28 November 2014

Abstract
Purpose Conjoined twins are a rare complication of 9 monozygotic twins and are associated with high perinatal mortality. Pygopagus are one of the rare types of conjoined twins with only a handful of cases reported in the literature.
Case summary We present the case of one-and-half month-old male pygopagus conjoined twins, who were joined together dorsally in lower lumbar and sacral region and had spina bifida and shared a single thecal sac with combined weight of 6.14 kg. Spinal cord was separated at the level of the conus followed by duraplasty. They had uneventful recovery with normal 15 months follow-up.
Conclusion Separation of conjoined twins is recommended in where this is feasible with the anticipated survival of both or one infant.

Keywords Pygopagus · Conjoined twins · Separation surgery

Introduction

Conjoined twins are identical twins (monozygotic and monochorionic) who develop from a single fertilized ovum in which incomplete embryonic division occurs at 13th to 15th day of conception. Its incidence ranges from 1:30,000 to 1:200,000 live births and 1 in 650–900 twin deliveries [1, 2]. Around 40–60 % are stillborn and approximately 35 % of live births expire within 24 h. Prognosis depends upon vitality of shared organs and congenital anomalies as well as type of twins. Girls predominate with 3:1 ratio [1]. Only 18–19 % of conjoint twins are joined at sacrum and are termed as Pygopagus twins. Few cases have been reported in literature with their successful surgical separations into two healthy individuals [2].

R. Awasthi · R. Iyengar (✉) · S. Rege · N. Jain
Department of Neurosurgery, Sri Aurobindo Medical College and PG Institute, Indore, India
e-mail: neuroraghu@yahoo.com

Case report

A one-and-half month-old male pygopagus conjoined twins (Fig. 1) was referred to our institute. Twins were delivered by cesarean section. They did not have any significant past neonatal history and were duly immunized.

Arbitrarily the twin on the right side was named as baby-1 and that on the left side was named baby-2. General condition of both the babies was normal. On examination, they were fused in the lower lumbar and sacral region (pygopagus) with combined weight of 6.14 kg. On examination, no motor or sensory deficit with no signs of hydrocephalus was observed. Rest neurological examination was unremarkable. Head, trunk, abdomen and all four limbs were separated. Genitalia and anal opening were normal. Magnetic resonance imaging (MRI) revealed that both were fused together dorsally in lower lumbar and sacral region and had spina bifida from fourth lumbar vertebra downwards and low-lying spinal cords tethered at fifth lumbar vertebra to first sacral vertebra (Fig. 2). Their filum terminale were fused together at second and third sacral vertebra in a single thecal sac. They had separate urinary bladders and rectums. There were no other shared organs or shared vascular anatomy.

Systemic examination of cardiopulmonary status did not reveal any significant abnormalities. Rest of the routine workup and blood investigations were within normal limits. Preoperative assessment and interdisciplinary communication, was vital to the success of the separation so that

Published online: 21 January 2015

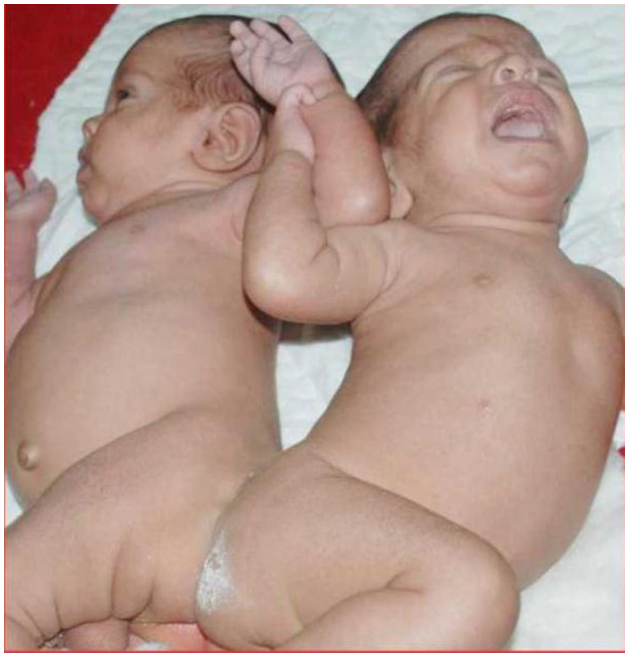

Fig. 1 Clinical picture of pygopagus conjoined twins

the planning for the surgery was started a day before with the department of anesthesia. Two teams of anesthetist were dedicated with the duplication of all the monitoring equipments. Planning for the postoperative period in the ICU was kept ready with two ventilators.

Both babies were positioned laterally on operating table and were draped properly to prevent hypothermia. Vertical incision was made over the attached spinal region. Care was taken during the exposure to avoid any neural injury. Exposure was extended one level above and one level below to allow dissection to extend from normal to abnormal anatomy. Under magnification, the dura was opened anteriorly. Adhered filum terminale was separated gently and two separable cords were observed in a single thecal sac. All non-neural and non-functional adhesive bands were transected. Adhesive bands were coagulated before cutting, to avoid bleeding. Spinal cord was separated at the level of the conus and duroplasty was done separately over both cords in a watertight fashion. Lumbar-dural fascia was mobilized and sutured in midline with 4-0 vicryl. Skin closer was done by mobilizing flaps with 4-0 nylon (Figs. 3, 4). Daily dressing was done and checked for any discharge from wound or any sign of flap necrosis. Suture removal was done on 14th postoperative day. Postoperatively both had an uneventful recovery with no neurological deficit (Fig. 5). Fifteen months follow-up was absolutely normal. From 10 months, both were crawling and from 14 months onward both can stand with support and walk with support. There were no sings of bowel or bladder involvement was noticed.

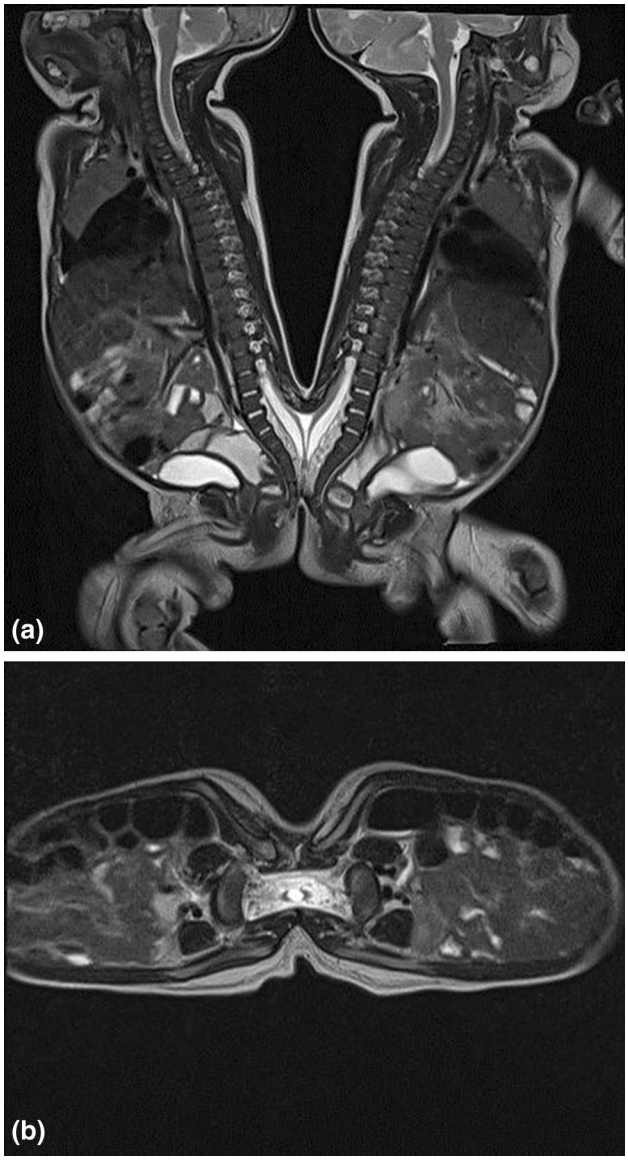

Fig. 2 MRI of pygopagus conjoined twins

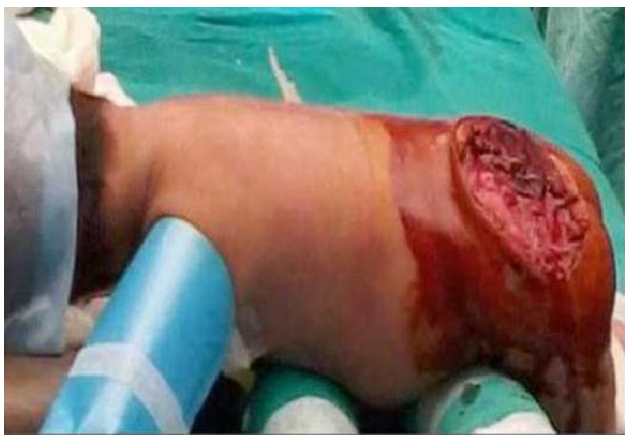

Fig. 3 Intra operative picture of one of the separated twin

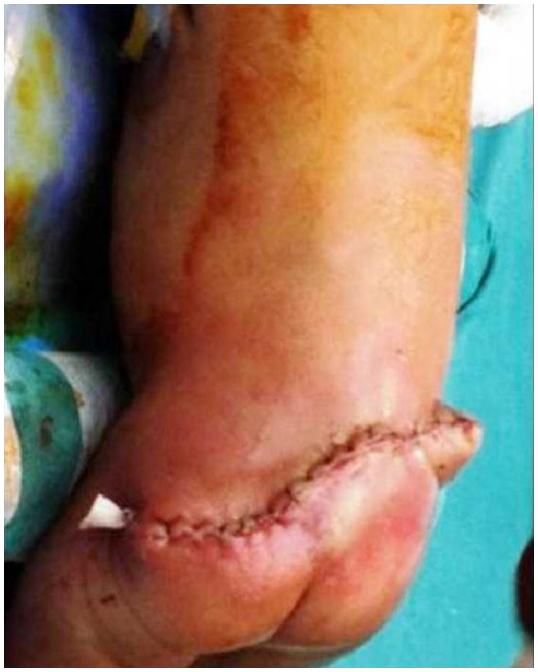

Fig. 4 Postoperative picture showing healthy suture line

Fig. 5 Separated and healthy twin babies

Discussion

Conjoined twins are always joined at homologous sites and the clinical classification is based on the most prominent site of union, combined with the suffix 'pagus' such as thoracopagus (chest), omphalopagus (umbilicus), ischiopagus (hip), pygopagus (rump), rachipagus (spine), craniopagus (cranium), cephalopagus (head), and parapagus (side) [3] (Table 1).

Table 1 Embryologic classification of conjoined twins [4]

Embryonic aspect	Type	Incidence (%)	Extent of union
Ventral (87 %)	–	–	–
Rostral (48 %)	Cephalopagus	11	Top to head to umbilicus
	Thoracopagus	19	Thorax, upper abdomen, conjoined heart
	Omphalopagus	18	Thorax, upper abdomen, separate hearts
Caudal (11 %)	Ischiopagus	11	Lower abdomen, genitourinary tract
Lateral	Parapagus	28	Pelvis, trunk, diprosopus 2 faces, dicephalus 2 heads
Dorsal (13 %)	Craniopagus	5	Cranial vault
	Rachipagus	2	Vertebral column
	Pygopagus	6	Sacrum

Two contradicting theories exist to explain the origins of conjoined twins. The older and traditional theory is fission, in which the fertilized egg splits partially and conjoined twins represent delayed separation of the embryonic mass after 12th day of fertilization. The second theory is fusion, in which a fertilized egg completely separates, but stem cells find alike stem cells on the other twin and fuse the twins together [1, 3].

The first successful surgical separation took place in 1689 in Switzerland by Johannes Fatio. One of the largest study at Red Cross War Memorial Children's Hospital in South Africa by Rode et al. [5] in 46 sets reported that pygopagus and ischiopagus twins have the best chances of long-term survival. Outcome in the surgery of conjoined twins is dependent on an experienced multidisciplinary team and the resources available only in a tertiary referral centre where the full range of medical, nursing and surgical specialties is present. Advances in diagnostic techniques particularly imaging modalities are helpful in planning for separation surgery [6, 7]. Inamdar et al. [1] suggested that the viability and prognosis of surgical separation of conjoined twins depend on the degree of union and number of shared organs.

Major factors for the successful outcome include mode of treatment (non-operative, emergency or elective procedure as per need), the standard approach, order of separation, the distribution of organs between the twins, meticulous aseptic surgical techniques, the reconstruction of divided organs and structures with tension-free primary closure of the wound [5].

In our case, both the twins have separated successfully from sacrum as both had separate spinal cord with common thecal sac and no vital organ was shared.

Conclusions

Improved survival rates for conjoined twins are due to advancement in perinatal and postnatal diagnostic techniques, meticulous surgical care, and careful follow-up offers the possibility of preserving or restoring normal function in many children. Separation is recommended in all cases where this is feasible with the anticipated survival of both or one infant. Psychosocial counseling of parents before surgery is one of the major important part.

Conflict of interest None of the authors has any potential conflict of interest.

References

1. Inamdar SA, Goel SS, Subhedar VS (2012) A thoracophagus conjoined twins with myelomeningocele: an unusual case. Int J Reprod Contracept Obstet Gynecol 1:61–63
2. Chatterjee SK, Debmaulik T, Talukder BC, Sen B, Sarangi BK (1986) Separation of pygopagus twins. Pediatr Surg Int 1:74–78
3. Osmanağaoğlu MA, Aran T, Güven S, Kart C, Ozdemir O, Bozkaya H (2011) Thoracopagus conjoined twins: a case report. ISRN Obstet Gynecol 2011:238360
4. Chitnis S, Derom C, Vlietinck R, Derom R, Monteiro J, Gregersen PK (1999) X chromosome-inactivation patterns confirm the late timing of monoamniotic-MZ twimming. Am J Hum Genet 65(2):570–571
5. Rode H, Fieggen AG, Brown RA, Cywes S, Davies MRQ, Hewitson JP et al (2006) Four decades of conjoined twins at Red Cross Children's Hospital—lessons learned. South Afr Med J 96:931–940
6. Spitz L (2003) Surgery for conjoined twins. Hunterian Lecture. Ann R Coll Surg Engl 85:230–235
7. Spitz L, Kiely EM (2003) Conjoined twins. JAMA 289:1307–1310