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Case Report

Thoracopagus conjoined twins: a case report

Rabi Narayan Satpathy*, Om Avishek Das, Sujata Swain, Amrita Nanda, Balaram Sahoo

Department of Obstetrics and Gynaecology, SCB Medical College, Cuttack, Odisha, India

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***Correspondence:**

Dr. Satpathy Rabi Narayan,

E-mail: drrabisatpathy@yahoo.com

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ABSTRACT

25 yr. old, G2P1 presented with premature labour pains at 33wks 3d of gestation and was referred to our tertiary centre as a suspected case of conjoined twin pregnancy based on a sonography report which revealed fetus with two heads. Decision in favour of LSCS was taken after counselling the patient and her attendants regarding the anomaly of the fetus and incompatibility of life and dangers of spontaneous vaginal delivery. LSCS was done with delivery of first twin by cephalic and second twin by breech extraction, both were preterm male babies joined anteriorly starting from thorax to umbilicus (Thoracopagus) with four arms and four legs, baby could not be revived and was declared clinically dead in few minutes by neonatologist. Photographs were taken and we tried to obtain consent for autopsy but attendants were reluctant. A review of the literature suggests that early diagnosis by a combination of ultrasound and MRI is essential to management, providing prognosis for viability and success of surgical separation and the opportunity for early counselling of parents and termination if indicated.

Keywords: Conjoint twin, Thoracopagus, Monoamniotic monochorionic

INTRODUCTION

Aristotle (384-322 B.C.) wrote about conjoined twins in his memoirs.¹ The term "Janiceps," indicating a form of conjoined twins, comes from the name Janus, a Roman god with two faces.² Conjoined twins called the "Biddenden Twins" survived for 30 years in Kent, England, around 1100 AD.³ In the Islamic history, Ibn Kathir mentioned that Hashim and Abd Shams, the sons of Abd Minaf Ibn Qusai were partially conjoined twins who were separated by their father.⁴ Overall, approximately 500 cases of conjoined twins are reported in the literature.^{5,6}

The most famous pair of conjoined twins was Chang and Eng Bunker (1811–1874), Thai brothers born in Siam, now Thailand. They travelled with P.T. Barnum's circus for many years and were billed as the Siamese Twins. Chang and Eng were joined by a band of flesh, cartilage, and their fused livers at the torso. Due to the brothers'

fame and the rarity of the condition, the term came to be used as a synonym for conjoined twins. They lived unseparated for 63 years and at the age of 31 they married two sisters who bore 21 children. They died within hours of each other, Chang from bronchitis and Eng from "fright" soon thereafter.⁷

Conjoined twinning occurs in 1 in 100 sets of monozygotic twins, 1 in 50,000 gestations or 1 in 250,000 live births.^{8,9} There is a greater incidence (70%) of female conjoined twin but the reason is unknown.¹⁰ Neither conjoined triplets nor recurrence of conjoined twins has been described. There is no mention in the literature of conjoined twins born to conjoined-twin parents.^{11,12} There are two theories explaining the aetiology of conjoined twins:

1. Collision theory by which embryonic axis fuse before tissue differentiation;

- Fission theory in which the embryonic tissue divides incompletely, remaining fused at some point or points.

The latter theory is more acceptable at about 2nd week in the normal twinning process, the inner cell mass splits into 2 separate and nearly equal halves each usually producing a single individual.¹³ Conjoined twins are monozygous and therefore will be of the same sex and usually with a monochorionic and monoamniotic placenta.¹⁴ Two cases of monochorionic, diamniotic conjoined twins have been reported in literature.¹⁵



Figure 1 & 2.



Figure 3 & 4.



Figure 5 & 6.

CASE REPORT

Pt P, 25 yr. old, G2P1 presented to us through LR with premature labour pain at 33wks 3d of gestation. She was referred from a district hospital to our tertiary teaching institution as a suspected case of conjoined twin

pregnancy based on a sonography report which revealed fetus with two heads. In her obstetric history she delivered a term female child vaginally one and half yrs. back, child alive and healthy, her second pregnancy continuing, LMP was on 8.11.12, she has taken IFA and calcium tablets, immunised with TT. No family h/o multiple pregnancy or congenital malformations, no consanguineous marriage or intake of any teratogenic drugs or exposure to any environmental toxins. It was a spontaneous conception and no h/o intake of any ovulation induction drugs nor any assisted reproductive technique. On examination average body built and nutrition with BMI 22.5, PR 86/min BP 110/60 mmHg, no pallor, icterus or oedema, systemic examination normal. P/A:- uterus over distended, fundal height corresponding to 36 wks., multiple fetal parts felt, exact presentation could not be made out, uterus relaxed, FHR(1) -146/min P/V :- Cx short soft, os 2cm dilated, presenting part high up. Investigations: - Blood gr- O+ve, Hb 11gm%, DC TLC Urine R&M, RBS within normal limits. She was advised for a repeat scan as the referral scan was inconclusive and after her scan in our department it was confirmed to be an intrauterine live conjoined twin with cephalic presentation with two heads, single thorax and abdomen, four lower limbs with two pelvis and a single heart with cardiac activity FHR 144/min, AGA 29wks 6d and 30wks 4d by femur length (FL) with EFW 2410 gms and AFI 12. Placenta single anterior not low-lying, Gr II maturity. Decision in favour of LSCS was taken after counselling the patient and her attendants regarding the anomaly of the fetus and incompatibility of life and dangers of spontaneous vaginal delivery. LSCS was done with delivery of first twin by cephalic and second twin by breech extraction, both were preterm male babies joined anteriorly starting from thorax to umbilicus (Thoracopagus) weighing 3 kg with four arms and four legs, babies did not cry after birth handed over to neonatologist but could not be revived and was declared clinically dead in few minutes, placenta with membranes delivered into in few minutes, it was a single placenta monoamniotic monochorionic type, no PPH, uterine shape normal, tubes ovary healthy. Photographs were taken and we tried to obtain consent for autopsy but attendants were reluctant. Post-operative period was uneventful and patient was discharged on eighth day.

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" meaning "that which is fixed". There are eight recognized configurations, as shown in Figure below: thoracopagus (chest), omphalopagus (umbilicus), pygopagus (rump), ischiopagus (hip), craniopagus (cranium), parapagus (side), cephalopagus (head), and rachipagus (spine).¹⁶ Conjoined twins can be further described as symmetrical or asymmetrical. Asymmetrical, or incomplete, conjoined twins result from

the demise of one twin with remnant structures attached to the complete twin, with the junction remaining at or near one of the common sites of union. Fetus-in-feto refers to asymmetrical monozygotic diamniotic intraparasitic twins.

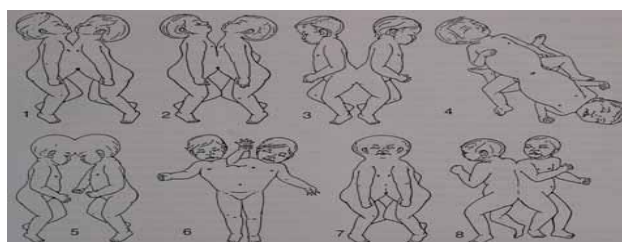


Figure 7: Different types of conjoined twins.

In above Figure 7, different types of conjoined twins described according to their site of conjunction:

- (1) Thoracopagus
- (2) Omphalopagus
- (3) Pygopagus
- (4) Ischiopagus tetrapus (four legs)
- (5) Craniopagus
- (6) Parapagus dipus (two legs)
- (7) Cephalopagus
- (8) Rachipagus.

Conjoint twins are generally classified three ways:¹⁷

1. 73 % are connected at mid torso (at the chest wall or upper abdomen)
2. 23 % at lower torso (sharing hips, legs or genitalia)
3. 4 % at upper torso (connected at the head)

Various classification systems have been proposed for this defect. Spencer classified conjoined twins on the basis of site of union i.e.: ventral or anterior union (cephalopagus, thoracopagus, omphalopagus, ischiopagus, and parapagus) and dorsal or posterior union (craniopagus, pyopagus, and rachipagus), while Potter and Craig simply classified on the basis of most common forms of twinning.^{16,18}

MANAGEMENT

Conjointment is seen as a medical condition that requires treatment, and the indicated treatment is invasive surgery to complete the process of splitting that was halted in the womb.

Surgical separation of nearly complete conjoined twins may be successful when organs essential for life are not shared. Consultation with a paediatric surgeon often facilitates parental decision making. It must be remembered that monozygotic twins are at increased risk to be discordant for structural malformations, most likely because the process of twinning is a teratogenic event which disturbs the timing of normal developmental processes. As a result, conjoined twins may have a

discordant structural anomaly that further complicates decisions about whether or not to continue the pregnancy.

Figure 8: Evaluation of conjoined twins.

System	Evaluation
Cardiorespiratory	Electrocardiogram (ECG) Echocardiography/Doppler ultrasound MRI/CT with contrast Angiogram
Alimentary tract	Contrast meal and enema Ultrasound Radioisotope scans (liver); technetium Tc99m-(Sn) colloid and excretion Tc99m mebrofenin Radioisotope scintigraphy
Genitourinary	Ultrasound Isotope renography Micturating cystourethrography Genitogram
Skeletal system	Radiography MRI (spinal cord)
Vascular	Doppler ultrasound Angiography
Cross-circulation	Radioisotope scan Tc99m-DMSA

Surgical separation of conjoined twins was attempted in 1950. The first successful pyopagus twin separation was performed in Louisiana in 1953. With the development of microsurgery and cardio-vascular surgery, many cases are successfully separated in different parts of the world.²¹ However, for ethical reasons, surgery should not be attempted if it deemed to be hazardous or if it may scarify or disable one twin for the sake of the others.²²

Successful surgical separation of conjoined twins needs multi-disciplinary approach of an experienced team [23]. The separation of conjoined twins has long been a surgical challenge. The timing of separation as a general principle is probably best to plan on an elective basis when infants are 9-12 months of age.²⁴

Sometimes emergency conditions such as intestinal obstruction, rupture of an omphalocele, congestive heart failure, obstructive uropathy and intractable respiratory embarrassment do occur and may necessitate emergency separation.²⁵ Sometimes critical condition of one twin may necessitate emergency separation.^{26,27}

The birth of two connected babies can be extremely traumatic and approximately 40–60 % of these babies are delivered stillborn with 35 % surviving just 1 day. The overall survival rate of conjoined twins is somewhere between 5 and 25 %. Over the years survival rates have improved as a result of more accurate imaging studies and better anaesthetic cooperative techniques.²⁸

REFERENCES

1. Barhmi R, Ferhati D, Melhouf MA, Nabil S, El Hamany Z, Kharbach A, et al. Jumeaux conjoints: Diagnostic échographique à 22 semaines. A propos d'un cas. *Rev Fr Gynécol Obstet.* 1996;91:547-50.
2. Roman era, approximately 170 AD Relics of Santa Mara.
3. Moussaoui RD. Jumeaux conjoints: A propos d'une observation. *Gynécologie Internationale.* 1997;6:59-61.
4. Kathir I, Gassick TL, Fareed M. *The Life of Prophet Muhammad: Al-Sira Al-Nabawiyya*, 1998.
5. Broussin B, Carles D. Les jumeaux conjoints: Diagnostic anténatal. *Med Foet Echo Gyn.* 1999;37:4-9.
6. Barth RA, Filly RA, Goldberg JD, Moore P, Silverman NH. Conjoined twins: Prenatal diagnosis and assessment of associated malformations. *Radiology.* 1990;177:201-7.
7. Wallace IW, Wallace A. *The Two*. New York, Simon and Schuster Inc, 1978.
8. Edmonds LD, Layde PM. Conjoined twins in the United States. *Teratology.* 1982;25:301 e8.
9. Spitz L. Conjoined twins. *Prenat Diagn* 2005;25:814e9.
10. Wilson RL, Cetrulo CL, Shaub MS. The prepartum diagnosis of conjoined twins by the use of diagnosis ultrasound. *Am J. Obstet Gynecol.* 1976;126:737.
11. Moussaoui RD. Jumeaux conjoints: A propos d'une observation. *Gynécologie Internationale.* 1997;6:59-61.
12. Puech F, Vaast P, Codaccioni X, Hubert D. Grossesses gemellaires et multiples. Etude anatomoclinique et prise en charge. Editions techniques-Encycl Med Chir (Paris-France) Obstetrique, 1993.
13. Wedberg R, Kaplan C, Leopold G, Porreco R, Resnik R, Benirschke K. Cephalothoracopagus (Janiceps) twinning. *Obstet Gynecol.* 1979;54:392-6.
14. Fitzgerald EJ, Toi A, Cochlin DL. Conjoined twins: Antenatal ultrasound diagnosis and a review of the literature. *J. Radiol.* 1985;58:1053-6.
15. Weston PJ, Ives EJ, Honors RLH, Lees GM, Sinclair DB, Schiff D. Monochorionic diamniotic minimally conjoined twins—a case report. *Am J. Med Genet.* 1990;37:558-61.
16. Spencer R. Anatomic description of conjoined twins: A plea for standardized terminology. *J Pediatr Surg.* 1996;3:941-4
17. Spencer R. Theoretical and analytical embryology of conjoined twins: part II: adjustments to union. *Clin Anat.* 2000;13:97e120.
18. O'Neill JA. *Conjoined Twins: in Pediatric Surgery.* O'Neill JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG. Mosby, 2006.
19. Miller AJW. Conjoined and parasitic Twins. Varma SK, Waalwyk K, Menahem S, Meagher S. First trimester diagnosis of conjoined twins aided by spatiotemporal image correlation. *J. Clin Ultrasound,* 2011.
20. Bega G, Wapner R, Lev-Toaff A, Kuhlman K. Diagnosis of conjoined twins at 10 weeks using three-dimensional ultrasound: a case report. *Ultrasound Obstet Gynecol.* 2000;16:388e90.
21. Kamak I, Cifteci AO, Buyukpcukcu N. Epigastric Heteropagus: a case report with review of the literature. *Eur J. Pediatr Surg.* 1999;9(5):347-50.
22. Groner JJ, Teske DW, Teich S. Dicephalus dipus dibrachius: an unusual case of conjoined twins. *J. Pediatr Surg.* 1996;31(12):698-700.
23. Al Rabeeah A. Conjoined Twins – past, present and future. *J. Pediatr Surg.* 2006;41(5):1000-4.
24. O'Neill JA, Holcomb HW, Schnauffer L, et al. Surgical experience with thirteen conjoined twins. *Ann Surg.* 1988;208:299-312.
25. Shapiro E, Ficker WR, Ternberg JL, Siegel MJ, Bell MJ, Manley CB. Ischiopagus tetrapus twins: Urological aspects of separation and 10 year follow up. *J. Urology.* 1991;145:120-5.
26. Dorig AT. Successful separation of ischiopagus tripus conjoined twins with one twin suffering from brain damage. *J. PeSiatr Surg.* 1993;28:965-8.
27. Stauffer JG. Conjoined twins. In *Neo natal Surgery*, 3rd edn. Eds Lister J, Irving IM, London, Butterworths. 1990;153-62.
28. Gore RM, Filly RA, Parer JT. Sonographic antepartum diagnosis of conjoined twins. It's impact on obstetric management. *JAMA.* 1982;247:3351-3.

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