International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Prajapati P et al. Int J Reprod Contracept Obstet Gynecol. 2023 Sep;12(9):2852-2855
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20232752

Case Report

A rare case of thoraco omphalopagus twins

Neel Prajapati, Farheenbanu Diwan*, Supriya Borole

Department of Obstetrics and Gynecology, Model Hospital, Bapu Nagar, ESIC, Ahmedabad, Gujarat, India

Received: 07 March 2023
Accepted: 02 August 2023

***Correspondence:**
Dr. Farheenbanu Diwan,
E-mail: farhindiwan27@gmail.com

ABSTRACT

Conjoined twins, popularly referred as Siamese twins, are twins joined in utero, affecting less than 1% of the monozygotic pairs. The frequency of conjoined twins has prevalence of 1.5 in 1 lakh births and thoracopagus is the most common. Early diagnosis is important as many patients opt for termination because of the grave prognosis. Though the prognosis of conjoined twin is generally low, there is limited evidence regarding the optimum method of termination of the pregnancy particularly in advanced gestational age. We hereby, report a rare case of successful medical termination of thoraco-omphalopagus twins at 23 weeks and 2 days of gestation. A 27-years old, 2nd gravid woman with previous vaginal delivery diagnosed with conjoined thoraco-omphalopagus twin pregnancy at 23 weeks and 2 days of gestation after the detailed ultrasonographic evaluation. Proper assessment and counselling were done regarding prognosis. Different options of termination were discussed and patient opted for medical termination of pregnancy with possibility of reverting to hysterotomy in case difficulty is encountered. After a 2 day of cervical preparation, a successful vaginal delivery was conducted. Even though this is an experience from a single case, medical termination can be safely preformed in carefully selected cases of conjoined twins beyond 20 weeks of gestation. The adequate cervical preparation, pain control and careful monitoring of the procedure are critical for optimal outcome.

Keywords: Conjoined twin, Thoraco-omphalopagus, Medical termination of pregnancy

INTRODUCTION

Conjoined twins, popularly referred as Siamese twins, are twins joined in utero. The most famous pair of conjoined twins was Chang and Eng Bunker (1811- 1874) brothers born in Siam (now Thailand). They travelled with P.T. Barnum circus for many years and were billed as Siamese twin. They were omphalopagus twins. Due to the brothers' fame and the rarity of the condition the term 'Siamese twins' came to be used as synonym for conjoined twins.[1]

Conjoined twins are entity, affecting less than 1% of the monozygotic pairs.[2] The frequency of conjoined twins has prevalence of 1.5 in 1 lakh births in which thoracopagus is the most (42%) common.[3] The other less common forms of conjoint twins include thoraco-omphalopagus (joined at chest and abdomen), pyopagus (joined at buttocks), ischiopagus (joined at the ischium) and craniopagus (joined at the head). Approximately 75% of the affected fetuses happen to be female.[4]

Due to high morbidity and mortality associated with conjoint twins' antenatal diagnosis is imperative. All the monozygotic twins should be screened for a possibility of conjointment. If conjointment is present then associated congenital anomalies like complex congenital heart diseases, lower GI anomalies like imperforate anus, genitourinary and CNS anomalies should be ruled out.[5]

We hereby, report a rare case of successful medical termination of pregnancy of thoraco-omphalopagus twins at 23 weeks and 2 days of gestation.

CASE REPORT

A 27-year-old, 2nd gravid women with previous normal vaginal delivery came to hospital for the 1st time for

routine ANC check up at 5 months of amenorrhea. The patient was normotensive with normal findings on general and obstetrics examination. She had no personal and family history of twins. Transabdominal sonography was performed as routine procedure and two fetuses at 23 weeks of gestation having two feal head at a fixed position facing each other and fused in midline suggestive of conjoin twins were noted. The patient was sent for further detailed anomaly scan. On 3D sonography, monochorionic monoamniotic live intrauterine conjoined thoraco-omphalopagus twins at 23+2 weeks of gestation with a possibility of agenesis of corpus callosum with ventriculomegaly and pericardial effusion with single truncus arteriosus in one of the twins were noted.

Figure 1: Ultrasonography of the fused twins at 22 weeks of gestation

Figure 2: Delivered conjoined twins at 22 weeks of gestation.

The assessment and counselling were done regarding the prognosis and the management. Patient opted for termination of pregnancy. The options for termination of pregnancy were discussed with the patient and she consented for medical termination of pregnancy with possibility of reverting to hysterotomy in case difficulty is encountered.

We performed cervical ripening with tablet mifepristone 200 mg given orally on day 1. On day 3, patient admitted and foley's induction was performed with foleys catheter no. 16 which was inflated with 30 CC of saline along with tablet misoprostol 200 mcg per vaginally kept. Patient was carefully monitored for progression and conjoined twins were successfully delivered vaginally approximately within 15 hours without any undue complication.

Patient recovered well and was discharged after 24 hrs. The follow up checkup after 1 week was uneventful.

DISCUSSION

Conjoined twins are rare. Its exact etiology is unknown but an incomplete division of the zygote between 13th to 15th day after fertilization can be the probable cause.[6] The overall survival rate for conjoined twins is approximately 25%.[7]

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.[8]

Table 1: Embryonic classification of conjoined twins.[9]

Embryonic aspect	Type	Incidence	Primordium	Extent of union	Separability
Ventral (87%)	-	-	-	-	-
Rostral (48%)	Cephalopagus	11%	Oropharyngeal membrane	Top of head to umbilicus	None
	Thoracopagus	19%	Heart	Thorax, upper abdomen, conjoined heart	Rare
	Omphalopagus	18%	Diaphragm	Thorax, upper abdomen, separate hearts	Likely 82% success
Caudal (11%)	Ischiopagus	11%	Cloacal membrane	Lower abdomen, genito-urinary tract	Likely 63% success
Lateral (28%)	Parapagus	28%	Cloacal membrane	Pelvis, variable trunk, diprosopus- 2 faces, dicephalus	Rare
Dorsal (13%)	Craniopagus	5 %	Cranial neuropore	Cranial vault	Unlikely without any sequalae
	Rachipagus	2%	Neural tube	Vertebral column	None reported
	Pygopagus	6%	Caudal neuropore	Sacrum	Likely 68% success

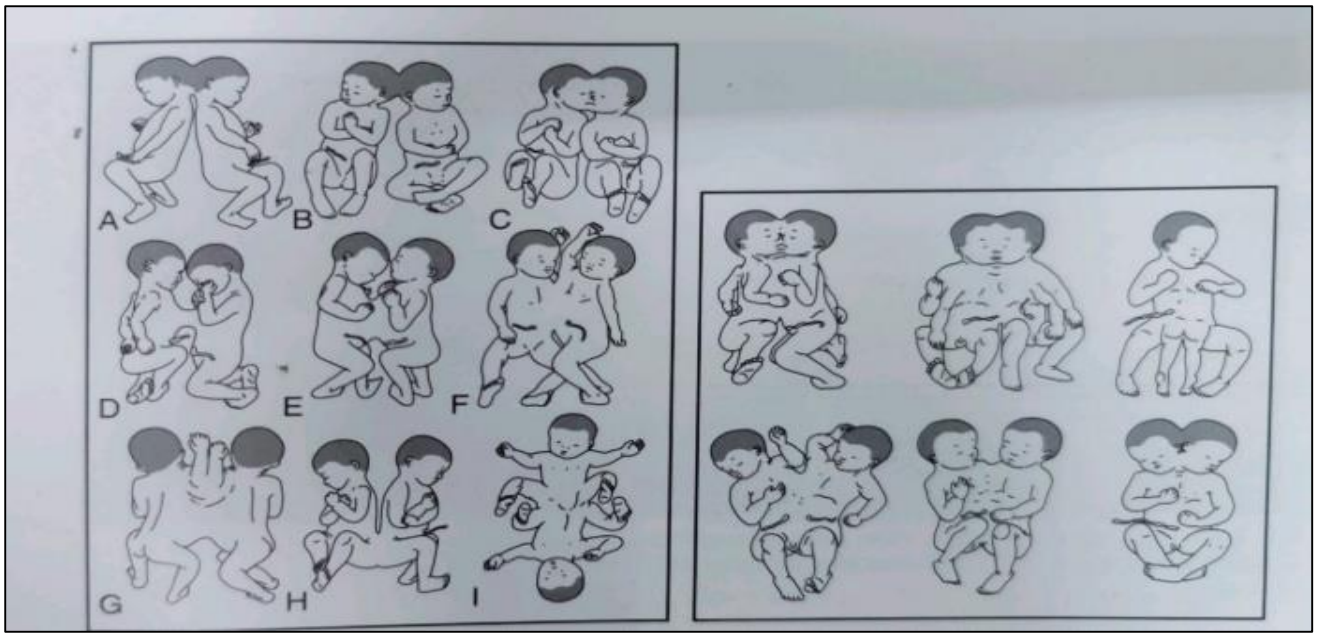

Figure 3: Differnet types of conjoined twins.

Conjoined twins can be identified using sonography in 1st trimester. This provides an opportunity for the parents to decide whether to continue ethe pregnancy. During sonographic interrogation, fetal poles are seen to be closely associated and does not change the relative position from one another. Other clues are more than 3 vessels in umbilical cord, fewer limbs than expected, spine hyperflexion bifid fetal pole and increased nuchal thickness. 3-dimensional ultrasound and color Doppler and MRI are valuable adjuncts to clarify the shared organs.[3]

Table 2: Ultrasonographic features of conjoined twins.[10]

S. no.	Ultrasonographic features of conjoined twins[10]
1	Bifid fetal pole in early pregnancy
2	Four vessel umbilical cord
3	Heads always at the same level
4	Relative position always constant
5	Extended spines

Early diagnosis followed by thorough counselling on likely prognosis is crucial for optimum management.[11,12] Approximate half are stillborn and one- third die within 24 hours of birth.[13] Many patients will opt for termination because of the grave prognosis.[3] However, in our case early diagnosis was not possible as the patient presented at second trimester of pregnancy. Other reports from developing countries also show the diagnosis of conjoined twins may be delayed until the third trimester or even up to the time of labor and delivery.[14,15]

Due to the lack of clear local guideline and experience in dealing with condition of this rarity, the exact methods for termination of pregnancy are unavailable. A literature review of methods of pregnancy termination for conjoint twins in second trimester revealed 75% delivered vaginally through medical induction while 18% underwent hysterotomy.[16] For pregnancy termination vaginal delivery is possible because the union is often pliable.[3] However, viable aged conjoined twins should be delivered by cesarean.

The decision for continuation or the termination of the pregnancy depends on the case concerned and should preferably individualized depending upon the facilities available regarding pediatric care with respecting the patient's decision.

CONCLUSION

In spite of the technological and surgical advancements, mortality of conjoined twins remains high. Also, the separation techniques are possible yet complicated and survival of the babies becomes difficult. When severe forms of conjoined twins are diagnosed prior to 24 weeks gestation, termination via vaginal delivery may be considered. The adequate cervical preparation, pain control and careful monitoring of the procedure are critical for optimal outcome.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Krishnan V. Obstetric 12 long cases, comprehensive case discussion, Chapter- twin pregnancy. 2010;281.
2. William obstetrics, 26th edition, chapter-multifetal gestation, conjoined twins. 2015;144.
3. Norton ME. Callen's ultrasonography in obstetrics and gynecology, book by, chapter- 48, conjoined twins. 2016.
4. Seo JW, Lee YS, Chi JG. Cross-sectional illustration on major types of conjoined twins. J Korean Med Sci. 1988;3(1):19-25
5. McMahon CJ, Spencer R. Congenital heart defects in conjoined twins: outcome after surgical separation of thoracopagus. Pediatr Cardiol. 2006;27(1):1-12
6. Schnaufer L. Conjoined twins, in Swenson's Pediatric Surgery. Raffensperger JG Ed. Appleton Century-Crofts, New York, NY, USA, 4th edition, 1980;910-20.
7. Abossolo T, Dancoisne P, Tuaillon J, Orvain E, Sommer JC, Rivière JP. Early prenatal diagnosis of asymmetric cephalothoracopagus twins. J Gynécol Obstétri Boil Reproduct. 1994;23(1):79-84.
8. Spencer R. Anatomic description of conjoined twins: A plea for standardized terminology. J Pediatr Surg. 1996;3:941-4.
9. Chitnis S, Derom C, Vlietinck R, Derom R, Monteiro J, Gregersen PK. X chromosome-inactivation patterns confirm the late timing of monoamniotic-MZ twimming. Am J Human Genetics. 1999;65(2):570-71.
10. Cunningham L. Williams Obstetrics, 23rd edition, McGrow Hill. 2009;39:871.
11. O'Brien P, Nugent M, Khalil A. Prenatal diagnosis and obstetric management. Semin Pediatr Surg. 2015;24(5):203-6.
12. Sen C, Çelik E, Vural A, Kepkep K. Antenatal diagnosis and prognosis of conjoined twins–a case report. J Perinat Med. 2003;31:427-30.
13. Carnevale FC, Marcus VB, Affonso BB, Pinto RP, Uenis T, João GM. Importance of angiographic study in preoperative planning of conjoined twins: case report. Clinics. 2006;61(2):167-70.
14. Gaym A, Berhan Y, Abadi GS, Wubishet T. Thoracopagus conjoint twins presenting as shoulder dystocia: a case report. Ethiop Med J. 2004;42:303.
15. Mohammad MA, Anyanwu LJC, Abdullahi LB. Management of conjoined twins in Kano, Nigeria: our experience and challenges in a low resource setting. Nigerian J Basic Clin Sci. 2018;15:92
16. Abubeker S. Successful dilation and evacuation for second trimester conjoined twin: a case report and review of the literature. J Med Case Rep. 2021;15:298.

Cite this article as: Prajapati N, Diwan F, Borole S. A rare case of thoraco-omphalopagus twins. Int J Reprod Contracept Obstet Gynecol 2023;12:2852-5.