

## CASE REPORT

### A RARE CASE OF THORACOPAGUS-CONJOINED TWINS WITH SINGLE HEART

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#### HOW TO CITE THIS ARTICLE:

N. Uma, D. Hemalathadevi, Ch. Sailaja, G. Rajalaxmi, Samyuktha. "A Rare Case of Thoracopagus-Conjoined Twins with Single Heart". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 28, April 06; Page: 4912-4915, DOI: 10.14260/jemds/2015/715

**ABSTRACT:** Conjoined twins is a rare disorder affecting one out of 30,000 to 1,00,000 births. A 25 year old unbooked multigravida of G3P2L1D1 with 8 months amenorrhoea with previous two L.S.C.S attended to our OPD with unknown LMP. Ultra sound revealed monochorionic, monoamniotic live conjoined twins of 28 weeks  $\pm$ 3 weeks with thoracopagus type contained single heart. The poor prognosis interms of perinatal morbidity & mortality was explained to the parents in such twins repeat elective L.S.C.S with bilateral tubectomy was done, delivered alive female conjoined twins with APGAR-7, with birth weight of 2.5kgs, babies fused at thorasic level, babies died after 5 days at new born care unit. Mother postoperative period was uneventful.

**KEYWORDS:** Conjoined twins, caesarean section, thoracopagus.

**INTRODUCTION:** An interesting anomaly unique to multiple pregnancies is conjoined twins. This is a rare disorder affecting 1 out of every 200 monozygotic twin pregnancies, 1 out of every 900 twin pregnancies, and 1 out of every 25,000 to 1,00,000 births.<sup>[1]</sup> Extremely on rare occasion division occurs after the embryonic disc and amniotic sac have formed and if the division of the embryonic disc is incomplete, conjoined twins result.<sup>[2]</sup> We report a rare case of conjoined twins who presented recently in our hospital.

**CASE REPORT:** A 25- year- old unbooked multi gravidia G<sub>3</sub>P<sub>2</sub>L<sub>1</sub>D<sub>1</sub> with 8 months amenorrhea presented to our OPD with unknown LMP. Her first pregnancy was full term emergency L.S.C.S. at government hospital for failed induction with pre eclamsia, delivered a live male child who died after 3 days, 3 years back. Her 2<sup>nd</sup> pregnancy was full term emergency L.S.C.S. At government hospital for CPD, delivered a live female child 2 years back, baby is alive and healthy. This is third pregnancy with no previous antenatal checkups. ML 5 years, she had only one menstrual cycle after last delivery. No family history of twins. The vital signs & general examination findings were within normal limits.

Per abdomen – Pfannenstiel scar was present. Height of uterus was 32 weeks size, multiple foetal parts palpable and more than three foetal poles felt. Uterus relaxed, no scar tenderness. On auscultation single foetal heart sound was heard with 154 beats per minute, regular.

Our clinical diagnosis was G<sub>3</sub>P<sub>2</sub>L<sub>1</sub>D<sub>1</sub> 32 weeks twin gestation with previous two caesarean sections.

**Investigations:** Hb: 10 gms, Blood Group: O positive, remaining blood investigations are within normal limits. Ultrasound showed monochorionic, mono amniotic live conjoined twins of 28 weeks+3weeks. thoracopagus type contained single heart with A-V septal defect with single umbilical cord contained two vessels with excess ammiotic fluid (AFI – 24).

MRI report showed thoracopagus conjoined twins with single heart.

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Provisional diagnosis was 25 years old G<sub>3</sub>P<sub>2</sub>L<sub>1</sub>D<sub>1</sub> with 28 weeks +3 weeks gestation with single heart conjoined twins with AV septal defect with polyhydramnios with previous two caesarean sections. We planned for termination by caesarean section after taking opinion of paediatric surgeon. Consent of parents was taken for L.S.C.S. with sterilization, after counselling the parents about the incompatibility of life of such twins in view of single heart and the risks and complications of vaginal delivery of conjoined twins in view of previous two L.S.C.S. Case was posted for repeat elective L.S.C.S. with bilateral tubectomy under spinal anaesthesia on 25/04/2014.

Per operative findings - both conjoined twins presented as vertex and extracted as such with little difficulty of delivery of second head. Delivered a live female conjoined twins with APGAR-7 with poor cry. Birth weight together 2.5 Kg. Both babies have separate heads with two upper and two lower limbs with single umbilical cord and fusion at thorax level. Cut end of umbilical cord showed single artery and vein.

Babies sent to new born care unit for further management. Twins are died after 5 days due to cardiac failure. Post-operative period of mother was uneventful. She was discharged from hospital on 8<sup>th</sup> P.O day.

**DISCUSSION:** Conjoined twins are mono ovular and have the same sex and karyotype. The phenomenon occurs predominantly in females. (3:1 female to male ratio). It results from incomplete division of the embryonic inner cell mass. The phenomenon happens early in gestation probably before the second week after fertilization. In the United States conjoined twins are commonly called as Siamese twins after Chang and Eng Bunker of Siam (Born in Thailand in 1811) who were displayed worldwide by P.T. Burnam. They were 63 years old when they died.<sup>[2]</sup>

Conjoined twins are classified according to the site of union that could be ventral in 87% of the cases and dorsal in 13% of the cases. Ventral unions include the following (Spencer 1996):-

- Thoracopagus (19%) - Joined at the chest.
- Omphalopagus (18%) - Joined at the anterior abdominal wall.
- Ischiopagus (11%) - Joined at the ischium.
- Craniopagus (11%) - Joined at the head.

**The distribution of dorsal unions is as follows:**

- Craniopagus (5%) - Joined at head.
- Pygopagus (6%) - Joined at the battuck.
- Rachiopagus (2%) - Joined at the spine.<sup>[1]</sup>

In our case it is Ventral union of thoracopagus with single heart.

Early diagnosis of conjoined twins are possible at first trimester by ultrasound examination.

A compressive ultrasound (TIFFA) examination between 18 and 22 weeks may be useful to determine the anatomy of the shared organs and to detect associated malformations which allow the parents to decide whether continue or not to continue the pregnancy.

The following ultrasound findings increase the probability that conjoined twins are present.

- The twins face each other.
- The heads are at the same level and plane.
- The thoracic cages are in unusual proximity.

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- Both foetal heads are hyper extended.
- There is no change in the relative position of the foetuses with movement or in repeat examination obtained hours or days later.<sup>[1]</sup>

Repeated 2D & 3D ultrasound examination, MRI scanning are useful for anatomical determination of cardiac connections between the twins.

The outcome of conjoined twins is poor. Approximately 40% of them will be still born, 35% will die within one day after delivery, and the overall survival rate of conjoined twins is between 5-25%.

**MANAGEMENT:** Consultation with paediatric surgeon often facilitates parental decision making. Termination of pregnancy is an option when the heart or brain is shared because in these cases attempt to separate usually fail. In our case separation is not feasible due to single heart. Once diagnosis is made plans should be made of caesarean delivery unless there are special circumstances indicating the possibility of safe vaginal delivery. Surgical separation of nearly complete conjoined twins may be successful when organs essential for life are not shared.<sup>[3]</sup> The only hope of independent life is through surgical separation. The first successful surgical separation of conjoined twins were achieved in 1953. Separation is feasible in Ischiopagus, Pygopagus, Omphalopagus, Parastic twins. Ideal time for separation at 9-12 months of age.

**CONCLUSION:** This is a rare disorder affecting one out of 200 monozygotic twins. The incidence of thoracopagus is 19%. TIFFA scan at 18-20 weeks helps to determine the shared organs and to detect malformation. Consultation with paediatric surgeon often facilitates parental decision making. In our case survival is not feasible due to single heart with A-V septal defect hence elective L.S.C.S. with sterilization done.

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**Fig. 1: MRI film showing conjoined twins**

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**Fig. 2: Photo showing Thoracopagus conjoined twins**



**Fig. 3: Photo showing conjoined twins with single umbilical cord**



**Fig. 4: Photo showing babies with two heads and two upper and lower limbs separately**

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### **FINANCIAL OR OTHER**

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Date of Submission: 11/03/2015.  
Date of Peer Review: 12/03/2015.  
Date of Acceptance: 25/03/2015.  
Date of Publishing: 06/04/2015.