CASE REPORT

Thoracopagus Conjoined Twin: A Case Report

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Introduction:

Conjoined twin is one of the rare congenital malformations. It occurs in approximately one in 50,000 to 100,000 live births, the most common type being thoracopagus-omphalopagus (including xipho-omphalopagus), and accounting for approximately 75% of this malformation[1]. It is usually associated with other anomalies. It is usually classified into complete or symmetrical and incomplete or asymmetrical sets. The imagings used for diagnosis are X-ray, ultrasonography, computed tomography (CT) scan, magnetic resonance imaging (MRI) of the abdomen and echocardiography[2]. The anatomical variability of both twins may jeopardize the successful surgical separation of conjoined twins. In fact, paediatric surgeons may be required to change their surgical approach and the planned dissection techniques used for separation in situ to achieve a good result. Thus, MRI is important when planning separation strategies for symmetrical conjoined twins. Here, a case of symmetrical twin of thoracopagus type with omphalocoele major is presented.

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Case report:

A conjoined twin of two female babies, attached at their thorax, was born at 37^{+} weeks of gestation to a 25 year old woman living in Dhaka city. It was the first pregnancy of the woman. The antenatal period had been uneventful. No drugs, illness, or radiation exposure during pregnancy were reported. There was no history of congenital anomalies in other children of the family or among other relatives. During pregnancy, the mother had an ultrasonography where the conjoined twin was identified. Delivary was done by caesarean section electively in a private hospital (Figure-1). After birth, the conjoined twin were managed by a paediatric surgeon and a neonatologist, and they were admitted to neonatology ward. The conjoined twin had a weight of 4000 gms. There was omphalocele major attached in lower part of the conjoined twin. The twining was attached at the thorax and upper abdomen. Both of them had separate head, extremities, and perineum. Examination of perineum showed female genitalia in both of them. Maternal

ultrasonography done before showed single heart with four chambers supplying the both members of the twin.

There was no respiratory problem after birth. Omphalocele major was managed initially with sterile vaseline gauge and gauge bandage coverings. Then an echocardiography was planned but their general condition was deteriorating gradually. So, they were shifted to neonatal intensive care unit (NICU) and the twin died on the fifth day of birth.

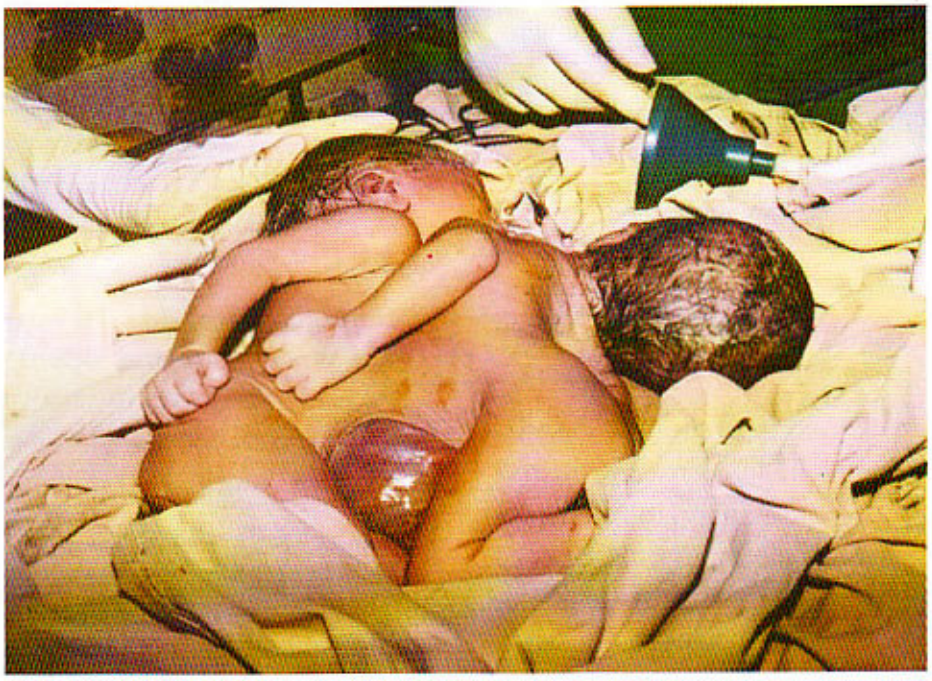

Figure-1: A conjoined twin delivered by elective caesarean section

Discussion:

Conjoined twin have high mortality. It was reported that intrauterine death was about 28% and death soon after birth was 54%, and the survival rate was only 18%[3]. Most common type is thoraco-omphalopagus. There are many theories on its embryonic formation. When monozygotic twining is expected to occur from germinal discs and if the division of the embryo is complete, they can develop into two identical persons. Otherwise, they would form complex fusion, developing into conjoined twins. Such twins are identical in sex and karyotyping[4]. Conjoined twins may develop from incomplete zygoting or by preimplantation embryonic fusion. In fact, asynchronus ovulation has been reported in mares. It is possible that the initially implanted embryo acts as a hindrance to the movement of the co-twin. It results in fixation of both embryos in the same uterine location[2].

Antenatal diagnosis (ultrasound, echocardiography, MRI, etc) can provide early information to diagnose, estimate associated anomalies and survival possibility. Thus, it can be planned accordingly to deal with after birth. Shi et al reported that they diagnosed most conjoined twins or other malformations after 18 weeks of gestation[5]. Mackenzie had

studied 14 sets of twin and reported that the earliest diagnosis time during antenatal period was ninth week of gestation, and at average at the twentieth week[3]. Here the case was diagnosed antenatally.

Conjoined twins may be recognized by following identifying points during gestational period: i) presence of breech presentation with indivisible trunk of twins by ultrasonography; ii) echocardiography indicating identical heart sound (thoracopagus); iii) alteration of facial position of foetus; iv) hyperextension of one or two sides of cervical vertebra; v) stretching of vault of the skull; vi) relative position stabilization of conjoint twins by serial scanning; and vii) single huge liver and heart[4].

Survival rate in delayed separation can be over 80-90%. The benefits lie in decreasing the risk of anaesthesia, giving time to find out unidentified anatomical malformation and planning procedures aimed to guarantee enough wound covering thereby reducing organic injury[1,3,4,6]. Shi et al observed that delayed separation done more than one month after birth in thorapagus-omphalopagus was safe and had high survival rate[5]. Emergency conditions that may force separation include one of the twins being difficult to resuscitate or it being stillborn, severe injury of conjoined parts during labor, associated severely distinctive anomalies, omphalocele rupturing, and one twin having incurable disease such as severe heart malformations[7].

Operation timing and separation methods vary according to types of anomalies and conditions of fused organs, associated malformations, and general conditions of infants. During separation, one should consider nature of bleeding, shortened operation time, assurance of the safety of the organic function, reconstruction of large defects, etc.

The application of anaesthetic and monitoring technique, advanced medical apparatus, artificial biosynthetic materials and cooperation of multidisciplinary teams improve the successful separation and survival rate.

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