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CONJOINED TWINS: A SYSTEMATIC REVIEW OF ANTENATAL DIAGNOSIS, OBSTETRIC MANAGEMENT AND PERINATAL OUTCOME WITH ILLUSTRATION OF A RARE UNDIAGNOSED THORACO-OMPHALOPAGUS TWIN PREGNANCY

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ABSTRACT Conjoined twins are a rare complication of monozygotic twinning associated with high perinatal morbidity and mortality. Advances in prenatal imaging have significantly improved early diagnosis; however, missed diagnoses continue to occur, especially in resource-limited settings. This systematic review summarizes contemporary evidence on the epidemiology, embryology, antenatal diagnosis, obstetric management and perinatal outcomes of conjoined twins. A comprehensive literature search was performed using major medical databases for studies published in the last decade. The available evidence suggests that thoraco-omphalopagus remains the most common type, ultrasonography is the primary diagnostic modality with fetal MRI serving as an important adjunct, and planned cesarean section is the preferred mode of delivery in most viable cases. Prognosis mainly depends on the extent of organ sharing, particularly cardiac and hepatic fusion. Despite technological advances, conjoined twins continue to present significant diagnostic and management challenges, emphasizing the need for a high index of suspicion in all monochorionic monoamniotic twin pregnancies.

KEYWORDS : Conjoined twins; Thoracopagus; Thoraco-omphalopagus; Antenatal diagnosis; Fetal MRI; Obstetric management

INTRODUCTION

Conjoined twins represent one of the rarest and most complex congenital anomalies encountered in obstetric practice. They result from abnormal monozygotic twinning, with an estimated incidence ranging from 1 in 50,000 to 1 in 100,000 live births and a marked female preponderance[1]. Most conjoined twins are stillborn or die shortly after birth, and only a small proportion survive long enough to be considered for surgical separation[2]. Thoracopagus and thoraco-omphalopagus twins constitute the majority of cases, accounting for nearly three-fourths of all conjoined twins[3].

Early antenatal diagnosis is crucial for appropriate counseling, pregnancy planning and postnatal management[4]. With the widespread use of high-resolution ultrasonography and fetal magnetic resonance imaging (MRI), diagnosis is now possible in the first trimester in many cases[5]. Nevertheless, missed or delayed diagnosis continues to be reported even in tertiary care centers, often resulting in unexpected intrapartum difficulties and increased maternal and fetal risk[6]. This article presents a systematic review of contemporary literature on conjoined twins, with illustration of a rare case of undiagnosed thoraco-omphalopagus twins encountered at term.

MATERIALS AND METHODS

This systematic review was conducted in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) 2020 guidelines. A comprehensive literature search was performed in PubMed, Scopus, and Google Scholar for studies published between January 2015 and December 2025. Search terms included: "conjoined twins", "thoracopagus", "thoraco-omphalopagus", "antenatal diagnosis", "fetal MRI", "obstetric management", and "perinatal outcome".

Two independent reviewers (Reviewer A & Reviewer B) screened titles and abstracts and performed full-text eligibility assessments. Any disagreements were resolved by consensus or third-party adjudication. Data extracted included study design, sample size, diagnostic modalities, obstetric management strategies, and outcomes.

A PRISMA flow diagram was used to document the identification, screening, eligibility, and inclusion of studies (see Figure 2). The PRISMA analysis ensured transparency of study selection and exclusion reasoning.

Figure 2 : PRISMA Flow Chart

Systematic Review of Literature

Epidemiology and Classification

Thoracopagus and thoraco-omphalopagus twins account for approximately 70–75% of all conjoined twins[7]. A consistent female predominance is observed, with a female-to-male ratio of approximately 3:1[1,8]. Based on the site of fusion, conjoined twins are classified as thoracopagus, omphalopagus, thoraco-omphalopagus, craniopagus, pygopagus, ischiopagus and rachipagus[2,3].

Embryology And Pathogenesis

Two mechanisms have been proposed: incomplete fission of the embryonic disc after the 13th day of fertilization and secondary fusion of two embryos[2,9]. Current embryological evidence favors the incomplete fission theory as the primary mechanism[2,9].

Antenatal Diagnosis

With present-day imaging, conjoined twins can often be diagnosed as early as 10–12 weeks of gestation by ultrasonography[5,10]. Characteristic features include persistent relative position of the fetuses, inability to separate fetal bodies, shared organs, a single umbilical cord and both fetal heads visualized in the same plane[4,5,10]. Fetal MRI serves as a valuable adjunct, particularly for detailed evaluation of shared cardiac, hepatic and gastrointestinal anatomy[11,12]. Despite these advances, missed diagnoses continue to be reported, especially in monochorionic monoamniotic twin pregnancies[6,13].

Obstetric Management And Mode Of Delivery

Management depends on gestational age at diagnosis, type and extent of fusion, presence of shared vital organs and parental wishes[7,14]. Early diagnosis allows consideration of termination in lethal or non-separable forms. In potentially viable cases, planned delivery in a tertiary care center with a multidisciplinary team is recommended[7,15]. Most authors advocate planned cesarean section for viable conjoined twins or advanced gestation[6,7,15]. Vaginal delivery is usually reserved for early gestation, intrauterine fetal demise or lethal anomalies[13,16]. Undiagnosed cases may result in obstructed labor or difficult intraoperative extraction with potential maternal and fetal morbidity[6].

Postnatal Outcome And Prognosis

Overall survival of conjoined twins remains poor, with most series reporting survival rates of approximately 20–30%[15,17]. Prognosis is primarily determined by the presence of shared vital organs, especially cardiac fusion, and the extent of hepatic and gastrointestinal sharing[17,18]. A shared heart is generally considered a contraindication to successful separation.

Illustrative case from our institution, Kasturba Hospital, Delhi.
A 30-year-old multigravida (G2P1L1) with a previous cesarean section presented in labour at term with a diagnosis of monochorionic monoamniotic twin pregnancy. Antenatal ultrasonography performed at 16 and 32 weeks failed to detect fetal conjunction. Both fetuses were in breech presentation, and cesarean section was planned.

During surgery, extraction of the first fetus was difficult and unexpected resistance was encountered while delivering the head. Careful intraoperative assessment revealed thoraco-omphalopagus conjoined twins. Both babies were delivered alive by breech extraction and cried immediately after birth. The combined birth weight was 4200 g. Postnatal imaging revealed a shared liver with separate cardiac and other visceral structures, making them potential candidates for surgical separation. This case highlights the persistent risk of missed antenatal diagnosis and the obstetric hazards associated with unexpected intrapartum discovery of conjoined twins.

DISCUSSION

Despite major advances in prenatal imaging, missed diagnosis of conjoined twins continues to be reported[6,13]. This is particularly relevant in monochorionic monoamniotic twin pregnancies, where failure to actively exclude conjunction can result in unexpected and hazardous intrapartum situations.

This case emphasizes the **critical role of early and meticulous ultrasonographic evaluation** in MCMA twin pregnancies for possible conjunction. Whenever suspicion arises, adjunctive imaging such as fetal MRI can provide superior soft-tissue delineation and help confirm shared organs. Regular training of sonographers, adherence to standardized protocols for twin pregnancy evaluation, and early referral to fetal medicine units can reduce the likelihood of missed diagnoses.

Additionally, routine anomaly scans performed later in gestation may focus on individual fetal biometry and viability rather than careful assessment of inter-fetal relationships. This underscores the importance of maintaining a high index of suspicion for conjunction in all MCMA pregnancies, particularly when fetal movements appear restricted or when both fetuses remain in a constant position relative to each other across serial scans.

CONCLUSION

Conjoined twins, although rare, represent a critical obstetric challenge associated with high perinatal risk. Early and accurate antenatal diagnosis is essential for optimal counseling and management, and every monochorionic monoamniotic twin pregnancy should be considered a potential conjoined twin pregnancy until proven otherwise.

Figure 1. Photograph of live-born thoraco-omphalopagus conjoined twins immediately after delivery.

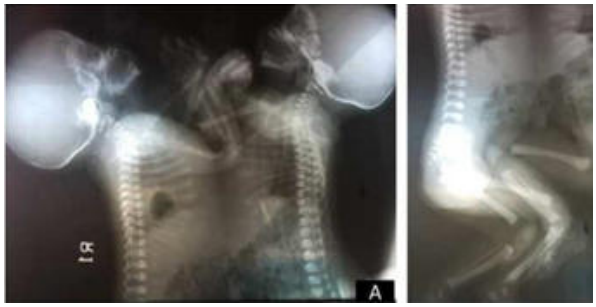

Figure 2. Plain radiograph (X-ray) of the thoraco-omphalopagus conjoined twins showing two distinct vertebral columns, two skulls, and fusion at the thoraco-abdominal region. The image demonstrates separate axial skeletons with close approximation and union in the thoracic and upper abdominal region, consistent with thoraco-omphalopagus configuration.

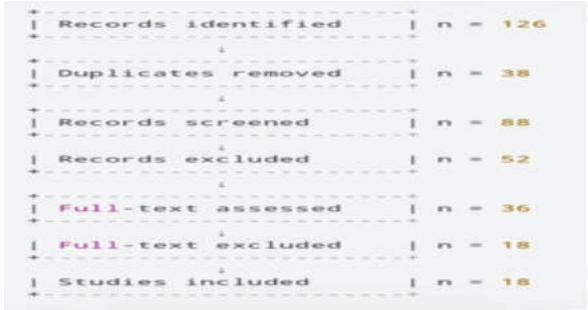

Figure 3: PRISMA 2020 Flow Diagram illustrating the study selection process for the systematic review of conjoined twins.

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