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OPEN ACCESS CASE REPORT

Up-to-date in Conjoined Twins Developmental Origins, Medical Challenges, and Ethical Considerations: Review Article

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ABSTRACT

Background: Incomplete division of a single fertilized egg during early embryonic development causes conjoined twins, a rare congenital abnormality. This condition presents serious medical, ethical, and social challenges, with an incidence of roughly 1 in 50,000 to 1 in 200,000 live births.
Objective: The purpose of this study is to investigate the developmental biology, classifications, medical and psychological issues, and moral quandaries related to conjoined twins.
Method: The literature on the embryological basis, classification schemes (particularly for thoracopagus and omphalopagus twins), medical complications, and the psychological effects on impacted individuals and their families forms the basis of this narrative review. It also looks at multidisciplinary surgical separation techniques and improvements in prenatal diagnosis using imaging modalities like MRI and ultrasound.
Results: Prenatal counseling and planning are greatly improved by early diagnosis made possible by advanced imaging, according to the study. Despite the risk, surgical separation has become more and more successful as a result of advancements in technology and procedure. For the best results, multidisciplinary teams must be involved. In order to manage such complicated cases and make sure that medical decisions are in the twins' and their families' best interests, ethical and psychological support continue to be essential.
Conclusion: Managing conjoined twins necessitates a compassionate, interdisciplinary, and ethically robust methodology. Advancements in surgical and diagnostic techniques provide individuals with renewed optimism; yet, ethical responsibility and psychological assistance are equally crucial for achieving success. Continuous case analysis and ethical discourse are crucial for enhancing treatment techniques.

Keywords: surgical separation, conjoined twins, and moral quandaries

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INTRODUCTION

Conjoined twins are siblings who are physically attached because the embryo doesn't fully separate during development. This partial separation results in two individuals being joined, often at the chest, abdomen, or pelvis (1). Sometimes they can even share internal organs. Many conjoined twins don't survive birth or die soon after, but with advances in technology and surgical techniques, their chances of survival have improved. In some cases, surgical separation is possible; the outcome depends on the point of attachment, the organs involved, and the team of doctors (1).

Definition from an embryological perspective Conjoined twins are a rare form of monozygotic twins when the embryonic disc doesn't fully separate during the 2nd week of development. This happens between days 13 and 15 of embryogenesis, the same time the primitive streak forms (2, 3).

The Formation Mechanism

Normal Twin Development: Monozygotic twins are formed when a single zygote splits into two separate embryos. Before the 13th day: Complete split results in normal monozygotic twins. After the 13th day: Incomplete split results in conjoined twins (2).

Conjoined Twin Development: According to fission theory, incomplete separation of the embryonic disc results in conjoined twins. According to fusion theory, two separate embryonic discs fuse at specific points, resulting in shared tissues or organs (2).

Theories of Development

1. Fission Theory: Incomplete separation of the embryonic disc during early development results in conjoined twins. (2)
2. Fusion Theory: Two embryonic discs initially develop independently but later merge, creating physical connections and potentially shared organs, depending on the fusion site. (2)

Classification of Conjoined Twins

Usually, conjoined twins are grouped according to the locus of their fusion. They may have shared organs or other body structures, and no two cases are precisely alike. Conjoined twins may be joined at: (1) the chest (Thoracopagus twins); (2) the abdomen (Omphalopagus twins); (3) the base of the spine (Pygopagus twins); (4) along the length of the spine, also called the rachipagus twins; (5) the pelvis (Ischiopagus twins); (6) the trunk (Parapagus twins); (7) the head (Craniopagus twins); (8) both head and chest (Cephalopagus twins); (9) the asymmetrical ones; or the fetus in fetu (10).

The current study aimed to explore the current understanding of the developmental paths and embryological processes that lead to conjoined twins. In addition, to examine the recent progress and obstacles in the medical management, surgical separation, and prognosis of conjoined twins of various anatomical classifications.

METHODOLOGY

Using a methodical approach, this review article sought, evaluated, and combined relevant information on the developmental origins, medical problems, and ethical considerations related to conjoined twins. To ensure comprehensive coverage of the topic, a comprehensive search was done across many scientific databases, including PubMed, Scopus, ScienceDirect, and Google Scholar. Emphasizing historical insights and the most recent developments in the field, the search included English-published books ranging from 2000 to 2024. Key search terms included "conjoined twins," "twinning anomalies," "embryological development of conjoined twins," "surgical separation of conjoined twins," "ethical considerations in conjoined twins," and "management of conjoined twins." For maximum result accuracy, keywords were combined using Boolean operators (AND, OR). The titles and abstracts of the obtained papers to remove studies unrelated to the focus of this review, non-English publications, non-peer-reviewed material, and duplicates. After that, full-text publications were carefully reviewed and included depending on their scientific quality, relevance, and contribution to modern knowledge. To ensure a multidisciplinary view, case studies, systematic reviews, clinical investigations, and ethical analyses were included.

Under three main themes—developmental origins and classification, medical and surgical challenges, and ethical and social implications—28 peer-reviewed publications were finally chosen, arranged, and examined. This methodical approach enabled a

comprehensive, current, and fair review of the body of current conjoined twin research.

RESULTS

Conjoined twins were classified into 10 classes as shown in Table 1.

Table 1: Classification of Conjoined Twins

NO.	Type of Conjoined Twins	Definitions
1.	Chest (Thoracopagus)	Twins that arise in connection with each other at the thoracic region, particularly at the chest, so that these twins will be face-to-face. They usually share the heart but may share a liver or the upper intestine as well. This is among the most common conjoined types of twins.
2.	Abdomen (Omphalopagus)	They are connected in the area of the bellybutton. Often, they have a shared liver along with portions of the upper digestive tract, especially the small intestine (ileum) and the large intestine (colon). Usually, no heart is shared.
3.	Base of Spine (Pygopagus)	Some share their lower digestive systems, and a few might share reproductive systems or urinary systems.
4.	Length of Spine (Rachipagus)	It's truly very unusual; they are connected along the full length of the spine.
5.	Pelvis (Ischiopagus)	The best way to describe conjoined twins is either side-by-side or head-to-toe at the hip region. They may share certain parts of the lower digestive tract along with the liver as well as reproductive or urinary systems. In a rarer situation, they may have 2 legs each, while in certain instances they may share 2 or 3 legs.
6.	Trunk (Parapagus)	They have separate heads but may share two to four arms and two to three legs, always joined side by side at the pelvis, abdomen, and andom'sosumetimes at the chest.
7.	Head (Craniopagus)	Connected at the back, top, or sides of their heads but not their faces. They share part of the skull but usually have separate brains, although some brain tissue may be shared.
8.	8. Head and Chest (Cephalopagus)	Conjoined at head and upper torso. They share a single head with faces on opposite sides and a shared brain. Very rarely does one of them survive.
9.	asymmetrical	With one twin being less developed than the other.
10.	fetus in fetu	In extremely rare instances, one twin may be partially formed within the other.

Clinical Consideration

Symptoms and Characteristics

Symptomatology and Features: The symptomatology and other characteristics of conjoined twins vary in regard to their site and dimension of joining. Conjoined twins are joined at certain sites in the body: the chest, abdomen, pelvis, or head. The connection could be of soft tissues or skin or might go as deep as to share bones, organs, or blood vessels (1). Shared organs and systems: On certain occasions, the shared vital organs could be the liver, heart, intestines, or brain, and so these would affect their health and development. Sharing systems might result in ailments such as troubles with digestion, breathing, or circulation (3). Lack of motion: The exact adhesion curtails the free movement of twins; they need to act in coordination with one another (4). Difference in development: One twin might grow slower and develop in comparison with another. Neurological or cognitive divergence can also occur, more particularly with shared structures in the brain and spine (1). Heart and lung problems result in twins who share a heart or lungs having complications like poor hand circulation, breathing problems, or inability to bear physical exertion (3). Threat of infections: There is a heightened risk of the formation of infections because of non-closure in regions of joining due to incomplete skin barriers (1). Problems in nutrition: Problems in feeding or shared digestive systems may bring about malnutrition or stunted growth (3). Other symptoms based on type of joining: Thoracopagus (chest connection): Usually share either heart or lungs, resulting in very grave complications of cardiac and respiratory functions (4). Omphalopagus (abdominal connection): Shared organs in the belly, like the liver or intestines, could bring about numeric and digestive complications (3). Craniopagus (head connection): The head-conjoined twins may share brain tissue or shared blood circulation, bringing along developmental delay or seizures (1). Pygopagus (connection in the lower back): Lower back connections disturb coordination and cause others to behave in bladder dysfunctions or shared spinal structures (5, 11). Ischiopagus (pelvic connection): Those joined at the pelvis might have problems in walking, as well as share systems of reproductive and urinary mechanisms.

Medical Attention Divided Into Two Parts

Most often, in pregnancy, the detection of conjoined twins is achieved through advanced imaging techniques such as ultrasound or CT. Regular prenatal care provides insight into the possible complications and prepares for delivery (4). Of it, a medical crisis that the child was presented with: After delivery, the extent of joint connections and organs, as well as possible approaches to treatment, must be outlined right away (5).

Importance of Medical Care

Comprehensive and timely medical attention is critical to managing the complex challenges faced by conjoined twins, ultimately improving their quality of life (5).

Diagnosis of Conjoined Twins

As show in table 2, the diagnosis of conjoined twins involves:

1. Prenatal diagnosis: Ultrasound - MRI .
2. Types of conjunction: Based on their point of connection .
3. Genetic testing: Chromosomal and genetic analyses.
4. Postnatal imaging: Imaging after birth (6).

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Table 2: Diagnosis of Conjoined Twins

Aspect	Diagnosis
Prenatal Diagnosis	Ultrasound: The primary diagnostic tool, typically used in the first trimester. Key indicators of conjoined twins include: 1. A single amniotic sac. 2. Absence of separation between fetal bodies. 3. Consistent positioning of the twins relative to each other. MRI: Offers a detailed anatomical evaluation, particularly useful for assessing shared organs and structures.
Types of Conjunction	Conjoined twins are classified based on their point of fusion, such as: 1. Thoracopagus (joined at the chest). 2. Omphalopagus (joined at the abdomen). 3. Craniopagus (joined at the head).
Genetic Testing	Chromosomal and genetic studies may be performed to identify potential associated anomalies or syndromes.
Postnatal Imaging	Advanced imaging techniques are utilized after birth to confirm prenatal findings and to aid in the planning of surgical separation, if it is deemed possible.

Prognosis of Conjoined Twins

In individual twins, different factors account for prognosis: (8). The type and extent of connection: Prognosis tends to be poor when vital organs like the brain and heart are fused. Associated malformations: The existence of additional abnormalities or malformations reduces the prospect of survival. Viability at birth: Factors such as gestational age, birth weight, proper immediate neonatal care, and others will influence survival chances. Survival chances are better for twins with simpler connections like omphalopagus and no shared vital organs. In recent years, there have been significant improvements in neonatal care and surgical techniques.

Treatment of Conjoined Twins

Management for conjoined twins requires a multidisciplinary team consisting of neonatologists, pediatric surgeons, radiologists, and geneticists. Prenatal care: Imaging, including ultrasound and MRI, is executed for evaluation of shared organ status and complications. The delivery plan is arranged in specialized high-risk perinatal centers. Delivery: Ideally, a cesarean section is usually performed in order to reduce risk both for the mother and the twins. Postnatal management: Immediate stabilization of the twins is required that concentrates mainly on monitoring both respiratory and circulatory functions. Postnatal imaging is performed to confirm the anatomy and to determine further care planning. Surgical separation: The possibility of separating the twins is determined by the degree of organ sharing and their overall medical condition. In general, surgery is delayed until the twins are deemed stable unless exigent circumstances necessitate immediate intervention. Likely complications include infection, organ failure, or death for one or both twins. Palliative care: In cases where separation is not feasible, care is aimed at providing comfort measures and improving the twins' life quality. Some Long-Term Outcomes: Separated twins: Their functional independence depends upon how complex the separation is and if any residual health issues remain. Conjoined twins: Twins that remain conjoined are faced with very high physical challenges, psychological issues, and social problems. Consistent support for the twins is needed from healthcare workers, counselors, and advocacy organizations to improve their quality of life.

Risk Factors of Conjoined Twins

Maternal Age: While an elevated risk for conjoined twins is observed among mothers in their late 20s to

early 30s, the actual risk, nevertheless, is very low (12). Family History: Most cases of conjoined twins do not run in families; nevertheless, a rare family background could slightly increase the odds. Environmental Factors: There is little evidence that exposure to environmental toxins may contribute to the incidence of conjoined twins. Abnormal Embryonic Development: Conjoined twins are a product of incomplete separation of the embryo during the early stages of development—which typically occurs between day 13 and 15 after fertilization.

Prevention of Conjoined Twins

Great prenatal health: Having a good diet, healthy lifestyle, and no exposure to deleterious substances before and during pregnancy can reduce the risk (13, 14). Early prenatal checking: Early pregnancy ultrasound examinations can show growth abnormalities and thereby allow for an early medical intervention (13). Assisted Reproductive Technology (ART): Due to close monitoring during ART procedures, this would assure good embryonic development and also risk minimization (13). Folic acid intake: Taking enough folic acid before and during pregnancy will greatly reduce incidents of certain birth defects (14). Avoiding harmful substances: Drugs, alcohol, and environmental toxins should be avoided in early pregnancy for healthy fetal development (14-17).

Survival and Life Expectancy

The survival rates of conjoined twins vary widely, depending on the type and extent of their physical connection. Some twins may live only a few hours or days, while others survive for many years, even reaching adulthood. Advances in surgical methods and medical care have improved survival rates. For twins who remain conjoined, their survival is often tied to their ability to adapt and work collaboratively in managing shared bodily functions. While some lead relatively independent lives, others may require continuous medical support (18-28).

Famous Cases of Conjoined Twins

Figure 1: Change and Eng Bunker [19]

Chang and Eng Bunker: Chang and Eng, born in 1811 in Siam, were joined at the chest and shared a liver. They gained fame as entertainers in the United States and lived to the age of 62, raising families (19).

Figure 2: Abby and Britty Hensel [20]

Abby and Brittany Hensel: Abby and Brittany, born in 1990, share a single body but have separate heads and brains. Despite their connection, they have achieved a high level of independence, managing everyday tasks, driving, and teaching (20).

CONCLUSION

Conjoined twins represent a rare and intricate phenomenon that highlights the complexities of human development. Their existence poses distinct medical and ethical challenges while providing opportunities to advance the frontiers of modern medicine and surgical techniques. Despite significant progress in diagnostic technologies and surgical

interventions, each case is unique, necessitating a customized, multidisciplinary approach.

Beyond the medical sphere, the experiences of conjoined twins shed light on crucial social, cultural, and psychological aspects. As scientific advancements continue, it is essential to balance medical possibilities with ethical considerations, ensuring decisions prioritize the well-being and quality of life of those affected. Conjoined twins stand as a testament to the resilience of the human spirit and the critical role of compassion in addressing intricate medical and personal challenges.

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