

DEXTROCARDIA: A COMPREHENSIVE REVIEW OF CONGENITAL HEART ANOMALY

Abstract

Dextrocardia, a rare congenital condition marked by the abnormal right-sided positioning of the heart within the chest cavity, challenges traditional cardiac anatomy. This comprehensive review explores the epidemiology, classification, investigations, and management of dextrocardia, emphasizing the need for early diagnosis and advancements in cardiac research. With an estimated occurrence of 1 in 10,000 live births, dextrocardia may manifest in isolation or coexist with congenital heart defects, classified as isolated dextrocardia, dextrocardia with situs solitus, or the more complex dextrocardia with situs inversus associated with Kartagener syndrome. Diagnostic investigations, including chest X-ray, electrocardiogram, echocardiography, and advanced imaging modalities like cardiac MRI and CT scans, play pivotal roles in identifying dextrocardia and evaluating its impact on cardiac function. Situs evaluation and genetic testing further contribute to a comprehensive understanding of associated anomalies. Management strategies vary based on the specific condition. Isolated dextrocardia often carries a favorable prognosis, while cases with congenital heart defects may require surgical intervention or cardiac catheterization procedures. Complex cases involving situs inversus demand a multidisciplinary approach, with coordination among pediatric specialists, gastroenterologists, and pulmonologists. Regular follow-up, education on healthy lifestyles, and collaborative efforts between healthcare professionals contribute to improved quality of life for individuals with dextrocardia. In conclusion, this review underscores the significance of early detection, thorough investigation, and collaborative management approaches in addressing the complexities of dextrocardia.

Authors

Dr. Natasha Verma

Assistant Professor
School of Health Sciences
Department of Physiotherapy
Garden City University
Bengaluru, Karnataka, India

Dr. S Senthil Kumar

Professor and Research Supervisor
School of Health Sciences
Department of Physiotherapy
Garden City University
Bengaluru, Karnataka, India

Sandhya Nagolu

Research Scholar
Saveetha Institute of Medical and Technical
Sciences
Saveetha University
Chennai, Tamil Nadu, India

Advanced imaging technologies and interdisciplinary cooperation are vital for optimal patient care, emphasizing the need for ongoing research to enhance our understanding of this rare congenital heart anomaly.

Keywords: Dextrocardia, situs, cardiac anatomy

I. INTRODUCTION

Dextrocardia is a rare congenital condition characterized by the abnormal positioning of the heart within the chest cavity. In this anomaly, the heart is flipped to the right side (dextrocardia) instead of its usual left-sided orientation (levocardia). This atypical cardiac anatomy results from an early developmental anomaly during embryogenesis. Dextrocardia can occur as an isolated finding or be associated with other congenital heart defects or syndromes. Understanding the epidemiology, classification, investigations, and management of dextrocardia is essential for early diagnosis, appropriate patient care, and advancements in cardiac research.

II. EPIDEMIOLOGY

Dextrocardia is considered a rare congenital heart defect, and its prevalence varies among different populations. The exact incidence is challenging to ascertain due to its rarity and the possibility of asymptomatic cases that go undiagnosed. However, studies estimate that dextrocardia occurs in approximately 1 in 10,000 live births. The condition affects both males and females equally, with no significant gender predilection.

The occurrence of dextrocardia can be classified based on the presence or absence of associated congenital heart defects or situs anomalies. Isolated dextrocardia refers to cases where the heart is the only organ affected. On the other hand, dextrocardia with situs solitus indicates the heart is reversed, but the other internal organs are in their normal left-sided orientation. Dextrocardia with situs inversus refers to a condition where both the heart and the other internal organs are mirror-image reversed.

III. CLASSIFICATION

- 1. Isolated Dextrocardia** is the simplest form of dextrocardia, where the heart is the only organ affected. It is typically not associated with other cardiac abnormalities or situs anomalies. Isolated dextrocardia may manifest with various subtypes, such as dextroposition, where the heart is displaced to the right side of the chest without significant cardiac malformation.
- 2. Dextrocardia with Situs Solitus:** In this type, the heart is positioned on the right side, but the other internal organs are normally left-sided. While the heart is reversed, the liver is on the right side, and the spleen is on the left side. This form of dextrocardia may also be associated with congenital heart defects, including ventricular septal defects (VSDs), atrial septal defects (ASDs), or anomalies of the great vessels.
- 3. Dextrocardia with Situs Inversus** is a more complex form of dextrocardia, where both the heart and other internal organs are mirror-image reversed. The heart is on the right side, the liver is on the left side, and the spleen is on the right side. Dextrocardia with situs inversus is often associated with Kartagener syndrome, a rare genetic disorder characterized by the triad of dextrocardia, situs inversus, and chronic sinusitis with bronchiectasis.

IV. INVESTIGATION

Diagnosing dextrocardia and conducting a comprehensive analysis of cardiac structures requires a combination of clinical evaluation, imaging studies, and diagnostic tests. Several investigations are commonly used to identify the condition and assess its impact on cardiac function and overall health.

- 1. Chest X-Ray:** An initial diagnostic step, a chest X-ray can reveal the abnormal positioning of the heart within the chest cavity. The characteristic "egg-on-side" appearance can be seen, indicating the presence of dextrocardia. Additional findings, such as cardiomegaly or other associated cardiac anomalies, may also be identified on the X-ray.
- 2. Electrocardiogram (ECG):** An ECG measures the heart's electrical activity and can help identify any associated cardiac abnormalities. It can also aid in differentiating between dextrocardia types based on the orientation of the electrical axis. In dextrocardia, the ECG leads will be reversed, and the QRS complex will be predominantly positive in lead a VR.
- 3. Echocardiography:** Echocardiography is a non-invasive imaging technique that uses ultrasound waves to create detailed images of the heart's structure and function. It helps assess the position of the heart, its chambers, valves, and blood flow patterns, providing valuable information about associated congenital heart defects. Echocardiography can also identify abnormalities in ventricular wall motion, valve regurgitation, and other cardiac functional issues.
- 4. Cardiac MRI and CT scan:** These advanced imaging modalities offer more detailed and three-dimensional images of the heart and surrounding structures. They can aid in identifying complex cardiac abnormalities, such as anomalous coronary artery origins, ventricular septal defects, or double-outlet right ventricles. Additionally, cardiac MRI and CT scans can provide comprehensive anatomical information, facilitating surgical planning in cases requiring intervention.
- 5. Situs Evaluation:** To determine the situs type (situs solitus or situs inversus), imaging studies such as abdominal ultrasound, CT scans, or MRI may be conducted to visualize the arrangement of other internal organs. Situs evaluation is essential for understanding the complete clinical picture and planning appropriate interventions.
- 6. Genetic Testing:** In cases where dextrocardia is suspected to be part of a genetic syndrome, genetic testing may be recommended to identify any underlying genetic mutations or abnormalities. In patients with Kartagener syndrome, genetic testing may reveal mutations in genes associated with ciliary dysmotility, such as DNAH5 or DNAI1.

V. MANAGEMENT

The management of dextrocardia depends on the individual's specific condition, including the presence of associated heart defects or other medical conditions. In isolated dextrocardia without any associated anomalies, the prognosis is generally favorable, and individuals can lead a normal and healthy life without significant cardiac limitations.

For cases with associated congenital heart defects, surgical intervention may be necessary to correct the structural abnormalities and optimize cardiac function. The timing and type of surgery will vary depending on the specific cardiac defect present. Cardiac catheterization procedures may also be employed for certain conditions, such as closing septal defects or treating anomalies of the great vessels.

In more complex cases of dextrocardia with situs inversus, the management may involve a multidisciplinary approach to address any associated organ anomalies. Coordinated care with pediatric specialists, gastroenterologists, and pulmonologists may be required to manage any complications associated with situs inversus and Kartagener syndrome.

Regular follow-up with a pediatric cardiologist or adult congenital heart disease specialist is essential to monitor the heart's function, overall health, and any potential complications. Patients with dextrocardia should be educated about the importance of maintaining a healthy lifestyle, including regular exercise and a heart-healthy diet, to support their cardiac well-being.

VI. CONCLUSION

Dextrocardia is a rare congenital heart defect characterized by the reversal of the heart's orientation within the chest cavity. The condition can manifest in various forms, from isolated dextrocardia to dextrocardia with situs solitus or situs inversus. Early diagnosis and comprehensive analysis of cardiac structures are crucial for appropriate patient management and care. Advanced imaging techniques, such as echocardiography, cardiac MRI, and CT scans, play a vital role in understanding the anatomy and function of the heart in dextrocardia cases. Collaborative efforts between pediatric cardiologists, cardiac surgeons, and other specialists are essential for providing optimal care and improving the quality of life for individuals with dextrocardia.

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