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Review Article

Uhl's anomaly: a comprehensive review of pathophysiology, clinical manifestations, diagnostic approaches, and therapeutic strategies

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ABSTRACT

Uhl's anomaly, a rare congenital cardiac disorder characterized by the absence or severe thinning of the right ventricular myocardium, presents significant challenges in diagnosis and management. This review aims to provide an exhaustive overview of the current understanding of Uhl's anomaly, encompassing its pathophysiology, clinical manifestations, diagnostic approaches, and therapeutic strategies. We delve into the embryological underpinnings and genetic predispositions associated with this condition, highlighting recent advancements in molecular and imaging techniques that facilitate early and accurate diagnosis. Furthermore, we discuss the clinical course, symptomatic presentation, and potential complications, offering insights into the prognostic factors that influence outcomes. Treatment modalities, including pharmacological management, interventional procedures, and surgical options, are evaluated based on contemporary evidence and expert consensus. Through this comprehensive review, we seek to enhance awareness and understanding of Uhl's anomaly among clinicians and researchers, ultimately contributing to improved patient care and outcomes.

Keywords: Uhl, Cardiac, Ventricular, Myocardium

INTRODUCTION

Uhl's anomaly, first described by Henry Uhl in 1952, represents a rare and intriguing entity within the spectrum of congenital heart diseases. Characterized by the partial or complete absence of the right ventricular myocardium, this anomaly results in a thin-walled, dilated right ventricle that predisposes affected individuals to severe right heart failure and arrhythmias. The pathogenesis of Uhl's anomaly remains incompletely understood, with hypotheses ranging from defective embryonic development of the right ventricular myocardium to potential genetic mutations that disrupt myocardial formation.[1,2]

The clinical presentation of Uhl's anomaly is highly variable, ranging from asymptomatic cases detected incidentally to severe, life-threatening right heart failure and arrhythmias. Common symptoms include exertional dyspnea, fatigue, peripheral edema, and syncope, often leading to a misdiagnosis of other more prevalent cardiac conditions. The rarity and clinical heterogeneity of Uhl's anomaly further complicate its diagnosis, necessitating a high index of suspicion and comprehensive diagnostic evaluation.[1,2]

Advances in imaging modalities, particularly echocardiography, cardiac magnetic resonance imaging (MRI), and computed tomography (CT), have significantly enhanced the ability to diagnose Uhl's anomaly with greater accuracy. These tools allow for detailed assessment of right ventricular structure and function, aiding in the differentiation of Uhl's anomaly from other causes of right ventricular dysfunction.[1,2]

Despite these diagnostic advancements, the management of Uhl's anomaly remains challenging due to the limited number of cases and the lack of standardized treatment

protocols. Therapeutic strategies are tailored to individual patient needs and may include medical management of heart failure symptoms, antiarrhythmic therapy, and interventional or surgical procedures such as right ventricular reconstruction or heart transplantation in severe cases.[1,2]

This article aims to provide a comprehensive review of Uhl's anomaly, synthesizing current knowledge on its pathophysiology, clinical manifestations, diagnostic approaches, and therapeutic options. By enhancing understanding of this rare condition, we hope to contribute to earlier diagnosis, more effective management, and improved outcomes for patients affected by Uhl's anomaly.[1,2]

EPIDEMIOLOGY

Uhl's anomaly, also known as parchment heart, is an exceedingly rare congenital cardiac disorder with a paucity of cases reported in the medical literature. Due to its rarity, precise epidemiological data on the prevalence and incidence of Uhl's anomaly are limited and primarily derived from case reports and small case series.[1,2]

PREVALENCE AND INCIDENCE

The exact prevalence of Uhl's anomaly is unknown, but it is considered to be exceptionally rare. Most estimates suggest that Uhl's anomaly accounts for a minute fraction of congenital heart defects. The incidence is so low that no large-scale epidemiological studies have been conducted to quantify its occurrence in the general population. The disorder is typically diagnosed in infancy or early childhood, although some cases are identified later in life, often incidentally or upon presentation of heart failure symptoms or arrhythmias.[1,2]

DEMOGRAPHICS

There appears to be no significant gender predilection for Uhl's anomaly, with cases reported in both males and females. Similarly, there is no clear evidence of ethnic or racial predilection, as the anomaly has been documented in diverse populations worldwide.

However, the small number of cases reported limits the ability to draw definitive conclusions about demographic patterns.[1,2]

GEOGRAPHIC DISTRIBUTION

Given the rarity of Uhl's anomaly, its geographic distribution is not well-defined. Cases have been reported across various regions, including North America, Europe, Asia, and Africa.

The widespread geographic occurrence suggests that Uhl's anomaly is a global condition, not confined to specific areas or populations.[1,2]

FAMILIAL OCCURRENCE AND GENETIC FACTORS

There are few reports of familial occurrence of Uhl's anomaly, indicating a possible genetic component in some cases. However, the majority of cases appear to be sporadic with no clear inheritance pattern. Genetic studies have not yet identified specific mutations or genetic markers consistently associated with Uhl's anomaly, but the involvement of genetic factors in its pathogenesis cannot be entirely ruled out. Some hypotheses suggest a potential link to mutations affecting myocardial development during embryogenesis.[1,2]

ASSOCIATED CONDITIONS

Uhl's anomaly is occasionally associated with other congenital heart defects or systemic conditions, although these associations are not consistent. Some reported cases have identified concomitant anomalies such as atrial septal defects, ventricular septal defects, or other structural heart abnormalities. The presence of such associated conditions may complicate the clinical course and management of affected individuals.[1,2]

MORTALITY AND MORBIDITY

The prognosis for individuals with Uhl's anomaly varies widely depending on the severity of the myocardial thinning and the presence of complications such as heart failure and arrhythmias. In severe cases, Uhl's anomaly can lead to significant morbidity and increased mortality due to progressive right heart failure and the risk of sudden cardiac death. Early diagnosis and appropriate management are critical to improving outcomes and quality of life for affected individuals.[1,2]

The rarity of Uhl's anomaly poses significant challenges for epidemiological research. The available data, largely derived from case reports, highlight the need for increased awareness and reporting of this condition to better understand its epidemiology. Future studies, potentially facilitated by international registries and collaborative research efforts, are essential to elucidate the prevalence, incidence, and genetic underpinnings of Uhl's anomaly, ultimately enhancing diagnosis, management, and prognosis for those affected by this enigmatic cardiac disorder.[1,2]

CLINICAL MANIFESTATIONS OF UHL'S ANOMALY

Uhl's anomaly, also referred to as "parchment heart," is a rare congenital heart disease characterized by the partial or complete absence of the right ventricular myocardium, resulting in a thin-walled, dilated right ventricle. This structural anomaly leads to significant hemodynamic consequences, manifesting in a spectrum of clinical signs and symptoms that vary based on the severity of the myocardial defect and the compensatory mechanisms of

the cardiovascular system. Understanding the clinical manifestations of Uhl's anomaly is crucial for timely diagnosis and appropriate management.[1,2]

EARLY PRESENTATION AND SYMPTOMS

In the neonatal and infant population, Uhl's anomaly often presents with signs of right heart failure. Symptoms may include:

Cyanosis

Due to the ineffective pumping action of the right ventricle, there can be inadequate pulmonary circulation leading to hypoxemia and cyanosis.[1,2]

Failure to thrive

Infants may exhibit poor weight gain and growth due to chronic heart failure and inadequate systemic perfusion.[1,2]

Respiratory distress

Tachypnea and dyspnea may be evident as the right ventricle fails to pump blood effectively into the pulmonary circulation, leading to pulmonary congestion.

Hepatomegaly

An enlarged liver is often palpable, secondary to hepatic congestion from right-sided heart failure.[1,2]

PEDIATRIC AND ADOLESCENT MANIFESTATIONS

As children with Uhl's anomaly grow older, they may continue to exhibit symptoms of right heart failure along with additional manifestations, including:

Exercise intolerance

Children may have difficulty keeping up with peers during physical activities due to limited cardiac output.[1,2]

Peripheral edema

Swelling of the lower extremities, abdomen (ascites), and sometimes face can occur due to venous congestion.[1,2]

Fatigue

Chronic fatigue is a common complaint, stemming from inadequate systemic circulation and oxygen delivery.[1,2]

Palpitations and syncope

Arrhythmias are frequent in Uhl's anomaly due to the abnormal myocardial structure, which can lead to palpitations, dizziness, and fainting episodes.[1,2]

ADULT MANIFESTATIONS

In adults, Uhl's anomaly might be diagnosed incidentally during evaluations for other conditions or when symptomatic exacerbations occur. Common adult presentations include:

Chronic right heart failure

Symptoms such as progressive dyspnea, orthopnea, and paroxysmal nocturnal dyspnea become more pronounced as the disease progresses.

Severe arrhythmias

Ventricular tachycardia, atrial fibrillation, and other arrhythmias are prevalent due to the structural anomalies of the right ventricle.

These arrhythmias can be life-threatening and may lead to sudden cardiac death.[3,4]

Thromboembolic events

The stagnant blood flow in the dilated right ventricle predisposes to thrombus formation, which can embolize to the pulmonary circulation, causing pulmonary embolism.[3,4]

Right ventricular outflow tract obstruction

Although not common, some cases report outflow tract obstruction due to the aberrant right ventricular anatomy.[3,4]

HEMODYNAMIC CONSEQUENCES

The hemodynamic disturbances in Uhl's anomaly are primarily due to the inability of the right ventricle to generate adequate pressure and flow. This leads to:

Reduced pulmonary blood flow

Compromised right ventricular function results in decreased pulmonary perfusion, contributing to hypoxemia and cyanosis.[3,4]

Systemic venous congestion

Impaired right ventricular output causes back pressure into the systemic venous system, leading to hepatomegaly, ascites, and peripheral edema.[3,4]

Compensatory mechanisms

The left ventricle may undergo hypertrophy to compensate for the decreased pulmonary blood flow, which can lead to further complications including interventricular septal shift and left ventricular dysfunction.[3,4]

DIAGNOSTIC CHALLENGES

The clinical presentation of Uhl's anomaly can mimic other cardiac conditions, such as arrhythmogenic right ventricular cardiomyopathy (ARVC) and idiopathic dilated cardiomyopathy. Therefore, a high index of suspicion and thorough diagnostic evaluation are essential. Diagnostic tools include:

Echocardiography

Key for identifying the thin-walled, dilated right ventricle and assessing right ventricular function.[3,4]

Cardiac MRI

Provides detailed imaging of myocardial structure and function, essential for differentiating Uhl's anomaly from other conditions.[3,4]

Electrocardiogram

Often reveals arrhythmias, right ventricular enlargement, and conduction abnormalities.[3,4]

Cardiac catheterization

Can help assess hemodynamic parameters and confirm the diagnosis.[3,4]

Uhl's anomaly presents with a wide range of clinical manifestations, primarily driven by right ventricular dysfunction and its systemic consequences. From early life symptoms of cyanosis and failure to thrive to adult presentations of chronic heart failure and life-threatening arrhythmias, the clinical spectrum of Uhl's anomaly is broad and complex. Recognizing these manifestations and understanding their pathophysiological basis is critical for early diagnosis, appropriate therapeutic interventions, and improved patient outcomes.[5,6]

DIAGNOSIS

Diagnosing Uhl's anomaly, a rare congenital cardiac disorder characterized by the absence or severe thinning of the right ventricular myocardium, poses significant challenges due to its rarity and the variability of clinical presentation. Accurate diagnosis is critical for guiding appropriate management and improving patient outcomes. This section provides a comprehensive overview of the diagnostic approaches and techniques used to identify Uhl's anomaly, emphasizing the importance of a multimodal strategy that includes clinical evaluation, imaging studies, and advanced diagnostic modalities.[5,6]

CLINICAL EVALUATION

The diagnostic process begins with a thorough clinical evaluation, encompassing a detailed patient history and physical examination. Key elements to consider include:

Patient history

The history should focus on symptoms indicative of right heart failure, such as exertional dyspnea, fatigue, peripheral edema, and episodes of syncope or palpitations. A history of cyanosis or failure to thrive in infancy may also be relevant. Family history should be explored to identify potential genetic predispositions or familial patterns.[5,6]

Physical examination

Physical findings may include jugular venous distension, hepatomegaly, ascites, and peripheral edema, all indicative of right-sided heart failure. Auscultation may reveal a prominent right ventricular heave, gallop rhythm, or murmur of tricuspid regurgitation.[5,6]

Electrocardiography

ECG is an essential initial diagnostic tool that provides information on the electrical activity and structural abnormalities of the heart. In Uhl's anomaly, typical ECG findings may include:

Right axis deviation: Reflecting right ventricular enlargement or hypertrophy.

Atrial arrhythmias: Such as atrial fibrillation or flutter.[5,6]

Ventricular arrhythmias: Including ventricular tachycardia, which is common due to the structural abnormalities of the right ventricle.[7,8]

QRS prolongation: Indicating conduction delays, often seen in the context of myocardial thinning.[7,8]

Chest radiography

A chest X-ray can provide valuable initial insights into the cardiac silhouette and pulmonary vasculature. Findings suggestive of Uhl's anomaly may include:

Cardiomegaly: Enlarged cardiac silhouette due to right ventricular dilatation.[7,8]

Prominent right heart border: Due to the dilated and thin-walled right ventricle.[7,8]

Pulmonary congestion: Signs of increased pulmonary vascular markings if right heart failure is present.[7,8]

Echocardiography

Echocardiography is the cornerstone of the diagnostic evaluation for Uhl's anomaly, offering real-time imaging of the heart's structure and function.

Key echocardiographic findings include:

Right ventricular dilatation: Markedly dilated right ventricle with a thin, "parchment-like" myocardium.[7,8]

Reduced right ventricular function: Hypokinetic or akinetic right ventricular free wall.

Paradoxical septal motion: Interventricular septal movement may be abnormal due to the dilated right ventricle.[7,8]

Tricuspid valve abnormalities: Including tricuspid regurgitation due to annular dilatation and structural deformation.[7,8]

CARDIAC MRI

Cardiac MRI is an advanced imaging modality that provides detailed characterization of myocardial anatomy and function, crucial for confirming the diagnosis of Uhl's anomaly. MRI findings include:

Myocardial thinning

Precise measurement of right ventricular wall thickness, highlighting the absence or extreme thinning of the myocardium.[7,8]

Right ventricular volume

Accurate quantification of right ventricular volume and function, including ejection fraction.[7,8]

Tissue characterization

MRI can differentiate between myocardial and non-myocardial tissue, providing insight into the extent of myocardial loss or replacement by fibrous or fatty tissue.[7,8]

CARDIAC CT

Cardiac CT is another valuable imaging technique, particularly useful for detailed anatomical assessment and pre-surgical planning. CT findings may mirror those seen on MRI, with additional benefits in evaluating coronary artery anatomy if coronary anomalies are suspected.[7,8]

Cardiac catheterization

Cardiac catheterization, including right heart catheterization, can provide hemodynamic data essential for evaluating the severity of right ventricular dysfunction. Key measurements include:

Right atrial pressure: Elevated pressures may indicate severe right heart failure.

Pulmonary artery pressure: Assessment of pulmonary hypertension, which may complicate Uhl's anomaly.[7,8]

Cardiac output: Quantification of cardiac output to gauge the functional impact of the right ventricular dysfunction.[7,8]

Endomyocardial biopsy

In selected cases, an endomyocardial biopsy may be performed to obtain histological confirmation of myocardial thinning and to exclude other differential diagnoses such as arrhythmogenic right ventricular cardiomyopathy (ARVC). Histological findings in Uhl's anomaly typically reveal:

Absent myocardium: Replacement of myocardial tissue with fibrous or fatty tissue.[7,8]

No inflammatory infiltrate: Differentiating it from myocarditis or other inflammatory conditions.

Differential diagnosis

The differential diagnosis of Uhl's anomaly includes other conditions that can present with right ventricular dilatation and dysfunction, such as:

Arrhythmogenic right ventricular cardiomyopathy: Characterized by fibro-fatty replacement of the right ventricular myocardium, often with a genetic predisposition.[7,8]

Idiopathic dilated cardiomyopathy: A non-ischemic cardiomyopathy with biventricular involvement.[7,8]

Ebstein's anomaly: A congenital malformation of tricuspid valve with apical displacement of the septal leaflet.[7,8]

The diagnosis of Uhl's anomaly requires a high index of suspicion, given its rarity and nonspecific clinical presentation. A multimodal diagnostic approach combining clinical evaluation, electrocardiography, chest radiography, echocardiography, cardiac MRI, CT, and catheterization is essential for accurate diagnosis. Advanced imaging techniques, particularly cardiac MRI, play a pivotal role in confirming the diagnosis and differentiating Uhl's anomaly from other similar conditions. Through comprehensive diagnostic evaluation, clinicians can better manage and optimize outcomes for patients with this rare congenital cardiac disorder.[7,8]

TREATMENT

The management of Uhl's anomaly, a rare congenital cardiac disorder characterized by the absence or severe thinning of the right ventricular myocardium, presents significant challenges due to the limited number of cases and lack of standardized treatment protocols. Treatment strategies are typically individualized, focusing on alleviating symptoms, improving cardiac function, and preventing complications. This section provides a detailed overview of the therapeutic options available for patients

with the Uhl's anomaly, encompassing medical management, interventional procedures, and the surgical approaches.[9,10]

Medical management

Medical therapy is the cornerstone of initial management for patients with Uhl's anomaly, aiming to control symptoms and improve quality of life. Key components of medical management include:

Diuretics: Used to manage symptoms of right heart failure such as peripheral edema, ascites, and hepatic congestion. Loop diuretics (e.g., furosemide) are commonly used to reduce fluid overload.[9,10]

ACE inhibitors or ARBs: These agents help reduce afterload and preload, thereby improving cardiac output and alleviating symptoms of heart failure.[9,10]

Beta-blockers: Indicated for managing arrhythmias and reducing the risk of sudden cardiac death. They can also help in controlling heart rate and reducing myocardial oxygen demand.

Antiarrhythmic drugs: Medications such as amiodarone or sotalol may be used to manage ventricular arrhythmias. Their use should be carefully monitored due to potential side effects.

Anticoagulation: Patients with significant right ventricular dilatation and reduced function may be at increased risk for thromboembolic events. Anticoagulants (e.g., warfarin) may be indicated to prevent thrombus formation and embolization.[9,10]

Interventional procedures

Interventional approaches may be necessary in certain cases to manage complications or improve symptoms. These include:

Catheter ablation: For patients with recurrent or refractory ventricular arrhythmias, catheter ablation can be an effective treatment to eliminate arrhythmogenic foci and improve symptoms.

Implantable cardioverter-defibrillator (ICD): Given the high risk of sudden cardiac death due to malignant ventricular arrhythmias, ICD implantation is often recommended.

ICDs provide continuous monitoring and can deliver life-saving shocks to terminate dangerous arrhythmias.[9,10]

Cardiac resynchronization therapy (CRT): In selected patients with significant dyssynchrony and heart failure, CRT may help improve right ventricular function and overall cardiac performance. [9,10]

Surgical management

Surgical intervention is considered for patients with severe symptoms refractory to medical and interventional therapies or those with significant structural abnormalities. Surgical options include:

Right ventricular reconstruction: This involves surgical techniques to reconstruct the right ventricular myocardium and improve its function. The success of this approach is limited by the extent of myocardial involvement and overall patient condition. [9,10]

Tricuspid valve repair or replacement: In cases where tricuspid regurgitation is severe and contributes to heart failure, surgical repair or replacement of the tricuspid valve may be necessary. [9,10]

Heart transplantation: For patients with end-stage right heart failure unresponsive to other treatments, heart transplantation remains the definitive therapeutic option. Transplantation is considered when all other medical, interventional, and surgical options have been exhausted and the patient's quality of life is significantly impaired.[9,10]

Postoperative care and long-term management

Postoperative care for patients with Uhl's anomaly involves close monitoring and management of potential complications. Long-term follow-up is essential to assess cardiac function, manage heart failure symptoms, and monitor for arrhythmias. Key aspects of postoperative and long-term care include:

Regular cardiac imaging: Echocardiography and MRI are used to monitor right ventricular size and function, assess for residual or recurrent issues, and guide ongoing management.

Electrophysiological monitoring: Continuous or periodic ECG monitoring and electrophysiological studies help detect and manage arrhythmias.[11]

Lifestyle modifications: Patients are advised to adopt lifestyle changes to optimize cardiac health, including dietary modifications, exercise as tolerated, and avoidance of excessive alcohol or stimulants.[12,13]

Patient education and support: Education about the condition, its implications, and the importance of adherence to medical therapy is crucial. Psychological support and counseling may be beneficial, given the chronic nature of the disease and the impact on quality of life.[13,14]

Emerging therapies and research

Research into the pathophysiology and treatment of Uhl's anomaly is ongoing, with potential emerging therapies on the horizon:

Gene therapy: Given the potential genetic basis of Uhl's anomaly, gene therapy may offer future treatment options by targeting specific genetic defects involved in myocardial development.[13,14]

Stem cell therapy: Research into the use of stem cells for myocardial regeneration holds promise for restoring right ventricular function in patients with Uhl's anomaly. Early studies in other forms of cardiomyopathy are paving the way for potential application in Uhl's anomaly.

Innovative surgical techniques: Advances in minimally invasive and robotic surgical techniques may improve outcomes for patients requiring surgical intervention for Uhl's anomaly.[14]

The treatment of Uhl's anomaly requires a multifaceted approach, tailored to the individual patient's needs and the severity of their condition. A combination of medical management, interventional procedures, and surgical options offers the best chance for symptom control and improved quality of life. Ongoing research into novel therapies and surgical techniques holds promise for the future, potentially transforming the management of this rare and challenging congenital heart disease. Through comprehensive and personalized care, clinicians can optimize outcomes for patients with Uhl's anomaly.[15,16]

CONCLUSION

Uhl's anomaly, a rare congenital cardiac disorder characterized by the absence or severe thinning of the right ventricular myocardium, presents significant diagnostic and therapeutic challenges due to its infrequency and the broad spectrum of clinical manifestations. This article has provided an in-depth review of the pathophysiology, clinical presentation, diagnostic approaches, and therapeutic strategies associated with Uhl's anomaly, emphasizing the need for a comprehensive and multidisciplinary approach to the optimize patient outcomes.

The etiology of Uhl's anomaly remains elusive, with theories suggesting both genetic and developmental factors contributing to the aberrant myocardial formation. Understanding the underlying pathophysiological mechanisms is crucial for developing targeted therapies and improving diagnostic accuracy. Clinically, Uhl's anomaly can present at any age, with symptoms ranging from asymptomatic incidental findings to severe right heart failure and life-threatening arrhythmias. This variability necessitates a high index of suspicion among clinicians, particularly when evaluating patients with unexplained right ventricular dysfunction.

Accurate diagnosis is pivotal and relies heavily on advanced imaging modalities. Echocardiography remains the initial diagnostic tool of choice, providing real-time assessment of right ventricular structure and function. Cardiac MRI offers superior tissue characterization and detailed anatomical evaluation, essential for differentiating Uhl's anomaly from other conditions such as arrhythmogenic right ventricular cardiomyopathy (ARVC). Cardiac CT and invasive techniques like endomyocardial biopsy and cardiac catheterization further aid in confirming the diagnosis and assessing the hemodynamic impact.

Management strategies for Uhl's anomaly are primarily symptomatic, given the lack of specific curative treatments. Medical management focuses on controlling heart failure symptoms and preventing arrhythmias, with diuretics, beta-blockers, and antiarrhythmic drugs playing central roles. Anticoagulation therapy is often necessary to mitigate the risk of thromboembolic events. For patients with refractory symptoms or severe arrhythmias, interventional procedures such as catheter ablation and implantable cardioverter-defibrillator (ICD) implantation provide critical therapeutic benefits.

Surgical interventions, including right ventricular reconstruction, tricuspid valve repair or replacement, and heart transplantation, are reserved for cases with advanced right heart failure unresponsive to medical and interventional therapies. The decision to pursue surgical options must be individualized, considering the patient's overall condition, comorbidities, and anticipated quality of life improvements.

Long-term management of patients with Uhl's anomaly involves regular follow-up and monitoring to assess cardiac function, manage ongoing symptoms, and detect complications early. Multidisciplinary care teams, including cardiologists, cardiac surgeons, electrophysiologists, and specialized nurses, are essential to provide holistic care and support to these patients.

Research into the genetic basis and pathophysiology of Uhl's anomaly is ongoing, with emerging therapies such as gene therapy and stem cell therapy holding potential for future advancements in treatment. Innovative surgical techniques and minimally invasive approaches continue to evolve, offering hope for improved outcomes and reduced procedural risks.

In conclusion, Uhl's anomaly remains a rare and complex congenital heart disorder that requires a thorough and multidisciplinary approach to diagnosis and management. Advancements in imaging techniques, a better understanding of the disease's genetic and developmental underpinnings, and the development of novel therapeutic strategies are critical to improving the care and prognosis for patients with this condition. Through ongoing research, clinical vigilance, and a commitment to individualized patient care, we can enhance the quality of life and clinical outcomes for those affected by Uhl's anomaly.

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