Research & Reviews: Journal of Medical and Health Sciences ISSN: 2319-9865

Pathogenesis and Pathophysiology of Hypoplastic Left Heart Syndrome

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Commentary

Received: 05-May-2023, Manuscript No. JMAHS-23-98122; **Editor assigned:** 09-May-2023, Pre QC No. JMAHS-23-98122(PQ); **Reviewed:** 23-May-2023, QC No. JMAHS-23-98122; **Revised:** 30-May-2023, Manuscript No. JMAHS-23-98122 (R); **Published:** 06-Jun-2023, DOI: 10.4172/2319-9865.12.2.008.

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Citation: Dufera M. Pathogenesis and Pathophysiology of Hypoplastic Left Heart Syndrome. RRJ Med Health Sci. 2023;12:008.

ABOUT THE STUDY

A rare congenital heart defect known as Hypoplastic Left Heart Syndrome (HLHS) causes the left side of the heart to be severely underdeveloped and unable to support the systemic circulation. It is thought to be responsible for 2 to 3 percent of all congenital heart disease. Poor feeding, cyanosis, and a decreased pulse in the extremities are some of the early symptoms. It is thought that a combination of genetic mutations and heart-related defects that alter blood flow are to blame for the multifactorial etiology. The affected structures include the left ventricle, aorta, mitral valve, and aortic valve, all of which can result in decreased systemic blood flow.

Using ultrasound or echocardiography shortly after birth, a diagnosis can be made prior to birth. The ductus arteriosus, a connection between the pulmonary artery and the aorta that closes shortly after birth, is the focus of initial treatment.

Pathogenesis

The majority of HLHS cases are sporadic, which means that they occur in people who have never had HLHS before. Since HLHS has been shown to be heritable and associated with particular gene mutations, some cases may have a genetic component. Intrauterine infarction, infectious changes, and selective left ventricular cardiomyopathy are all potential contributors.

Altered blood flow

According to a well-known theory known as the "no flow, no grow" hypothesis, primary defects in the aortic and mitral valves result in malformations of the left ventricle and its outflow tract. These primary defects can be divided into those that result in reduced left ventricular filling or obstruction of the outflow tract. Obstruction of the outflow tract causes left ventricular hypertrophy and a narrowing of the lumen. Aortic stenosis provides one illustration of this. The left ventricle experiences additional stress in utero as a result of aortic stenosis that develops during fetal development. This may eventually result in decreased left ventricle perfusion, which is thought to impede ventricular growth.

Pathophysiology

The ductus arteriosus is still open at birth, and the lungs have a higher-than-average resistance to blood flow. This takes into consideration sufficient oxygenation through blending between the atria and an ordinary appearance upon entering the world. Blood flow through the ductus is restricted and flow to the lungs is increased when the ductus begins to close and pulmonary vascular resistance decreases. The left side of the heart pumps oxygen-rich blood to the rest of the body in accordance with typical anatomy. A number of cardiac anomalies can occur in HLHS patients, eventually resulting in a small left ventricle that is unable to provide sufficient blood flow to the rest of the body. The disease can be classified into three main anatomic variants based on whether the aortic and mitral valves are stenotic or atresiastic.

The most severe form of HLHS is called Mitral Atresia and Aortic Atresia (MA-AA). In this condition, there is no blood flow to the left ventricle, so there is no chance of left ventricular output. In the Mitral Stenosis and Aortic Atresia (MS-AA) subtype blood can fill the left ventricle, but it can't be provided to the fundamental flow by means of the hypoplastic rising aorta. The Mitral Stenosis and Aortic Stenosis (MS-AS) subtype is the mildest form. Even though the left ventricle is able to provide some blood flow to the rest of the body in these patients, the left ventricular systemic output is still insufficient overall.

Diagnosis

Echocardiography can be used to diagnose hypoplastic left heart syndrome either before or after birth. Normal discoveries incorporate a left ventricle and aorta, irregularities of the mitral and aortic valves, retrograde stream in the cross over curve of the aorta, and left-to-right stream between the atria. It is frequently detected in the second trimester of pregnancy, between the ages of 18 and 24.