Rheology of discrete subaortic stenosis

A M Cilliers, M Gewillig

The discrete form of subaortic stenosis is thought to be an acquired lesion, the aetiology of which may be a combination of factors which include an underlying genetic predisposition, turbulence in the left ventricular outflow tract, and various geometric and anatomical variations of the left ventricular outflow tract. A review of hypotheses relating to its aetiology is provided.

RECENT AETIOLOGICAL THEORIES

The following series of hypotheses on the aetiology of this lesion were preceded by earlier observations by Rosenquist and colleagues and Somerville and associates, who concluded that fixed subaortic stenosis is a lesion that is acquired because of a flow disturbance in the left ventricular outflow tract.

Turbulence theory

The fact that discrete subaortic stenosis can recur after surgical removal led to the supposition that the lesion occurs as a result of a pathological process that was left unaltered by the surgery. An echocardiographic study by Gewillig and colleagues in 1992 showed that abnormal flow patterns are present in patients with discrete subaortic stenosis and that chronic flow disturbances are the cause of the development of the stenosis and its recurrence. Causes of chronic flow disturbances that may stimulate the endothelium to undergo transformation are mainly anatomical. They are as follows:

- apically situated muscular ventricular bands that reach the outflow tract, causing disturbance of flow in the subaortic area
- a septal ridge, which is an offshoot of a muscular band situated more apically in the outflow tract, causing turbulence that reaches the subaortic region
- malalignment of the interventricular septum, resulting in protrusion of the septum into the left ventricular outflow tract, so causing flow disturbances
- a long left ventricular outflow tract associated with increased mitral–aortic separation, resulting in an enhanced flow disturbance in the left ventricular outflow tract.

Geometric theory

In 1987, Zielinsky and colleagues found that most patients who developed subaortic stenosis had a malaligned VSD with anterior deviation of the infundibular septum, while Rosenquist and associates showed a twofold increase in mitral–aortic separation compared with normal hearts. In 1993, Kleinert and Geva confirmed the above findings but also showed that there was exaggerated aortic override in patients with subaortic stenosis and an intact interventricular septum. An aortoseptal angle (defined as the angle formed by the long axis of the ascending aorta and the plane of the ventricular septum) of < 130° was a prominent feature in their group of patients with subaortic stenosis (fig 1). The steeper angle results in a flow disturbance in the left ventricular outflow tract. The turbulent flow produced by the angle may induce an abnormal...
The pathophysiology of discrete, fixed subaortic stenosis is now closer to being understood. The definitive answer may evolve with further cellular biological research. Studies on endothelial and other growth factors may eventually provide the means of preventing and treating this intriguing heart lesion.

CONCLUSIONS

UNUSUAL ASSOCIATIONS WITH DISCRETE SUBAORTIC STENOSIS

Bilateral fibrous ridges have been noted in patients with a doubly committed VSD. Turbulence is maximal in the area of the subpulmonary and subaortic ridges, and the histopathological similarities suggest a common mechanism for the development of fibrous ridges in patients with this type of VSD. The increased flow across the left ventricular outflow tract in patients with a patent arterial duct may be the stimulant for membrane development, while the haemodynamic changes accompanying pulmonary artery banding have been implicated in the development of subaortic stenosis related to a secondary leftward shift of the conal septum. A further intriguing association is the trilogy of double chambered right ventricle with subaortic narrowing. The severity or progression of the subpulmonary obstruction and any relation to the severity of subaortic narrowing is unclear.

CONCLUSIONS

The pathophysiology of discrete, fixed subaortic stenosis is now closer to being understood. The definitive answer may evolve with further cellular biological research. Studies on endothelial and other growth factors may eventually provide the means of preventing and treating this intriguing heart lesion.

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REFERENCES


Figure 1 An illustration of the long axis of the heart showing the aortopulmonary angle between the long axis of the aortic root and the proximal ascending aorta and the midline of the interventricular septum. An angle of < 130° is thought to contribute to increased turbulence in the subaortic area, resulting in the development of a subaortic ridge. Drawing reproduced from Kleinart and Geva with permission.
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