

Tunnel type left ventricular outflow tract obstruction: An unusual complex congenital cardiac anomaly in adult

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A 24-year-old man, referred to our center with generalized body edema and hypotension. He had a complex congenital heart disease that finally discharged after konno operation.

Introduction

Congenital aortic stenosis is categorized as subvalvular, valvular and supra-valvular forms. Aside from coarctation of aorta, supra-valvular obstruction is an extremely uncommon entity in adults. Two other types of subvalvular aortic stenosis are discrete form and the fibromuscular type. Together, these lesions account for less than 20% of all cases of left ventricular outflow obstruction in children and both are uncommon in adult patients. Association between discrete subaortic stenosis and other sub aortic anomalies is a well known but rarely reported occurrence.¹ Fixed subaortic stenosis may be due to a discrete fibrous membrane, a muscular narrowing, or a combination of the both. The obstruction may be focal, as a discrete membrane, or more diffuse, resulting in a tunnel leading to left ventricle. The discrete form of fibromuscular subaortic stenosis is most frequently encountered (90%),² but tunnel type lesions are associated with a greater degree of stenosis. The prevalence of discrete sub aortic stenosis in adult with congenital

heart disease is 6.5%.² A bicuspid aortic valve is present in 23% of patients.³ When a tunnel type of subaortic stenosis coexists with hypoplasia and narrowing of the LV aortic junction, aortoventriculoplasty (Konno operation) is the reasonable procedure using the modification described by Misbach and Ebert and others.⁴⁻⁶

Case Report

A 24 year-old man, with echocardiographic diagnosis of tunnel type left ventricular outflow tract obstruction (Fig. 1), bicuspid aortic valve (Fig. 2), and severe valvular aortic stenosis was admitted with clinical diagnosis of biventricular failure.

The patient had previously undergone stenting of coarctation of aorta. Left cardiac catheterization and echocardiography revealed normal coronary angiography, bicuspid aortic valve, with severe aortic stenosis, tunnel type LVOT obstruction and severe mitral regurgitation.

Concerning the patient's echocardiography findings and his clinical situation, he underwent a successful Konno operation (Fig. 3) and mitral valve repair using annuloplasty ring, artificial chordae and cusp repair. Ten days after

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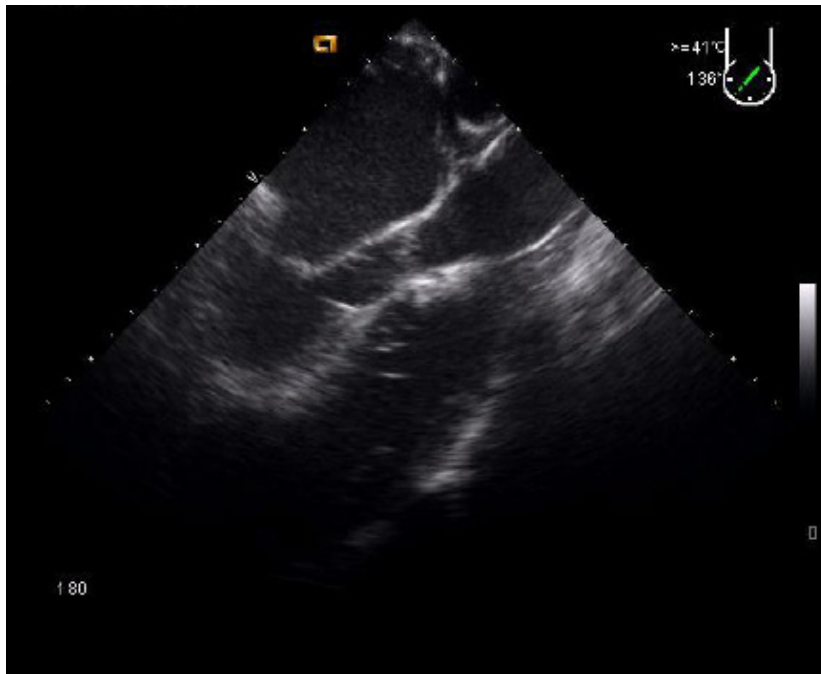


Figure 1: Transesophageal echocardiography shows tunnel type left ventricular out flow tract and subaortic web

operation transesophageal echocardiography revealed normally functioning prosthetic aortic valve and no evidence of subaortic stenosis,

with no significant mitral regurgitation. The patient was discharged. During the follow up period he is well and free of symptoms.

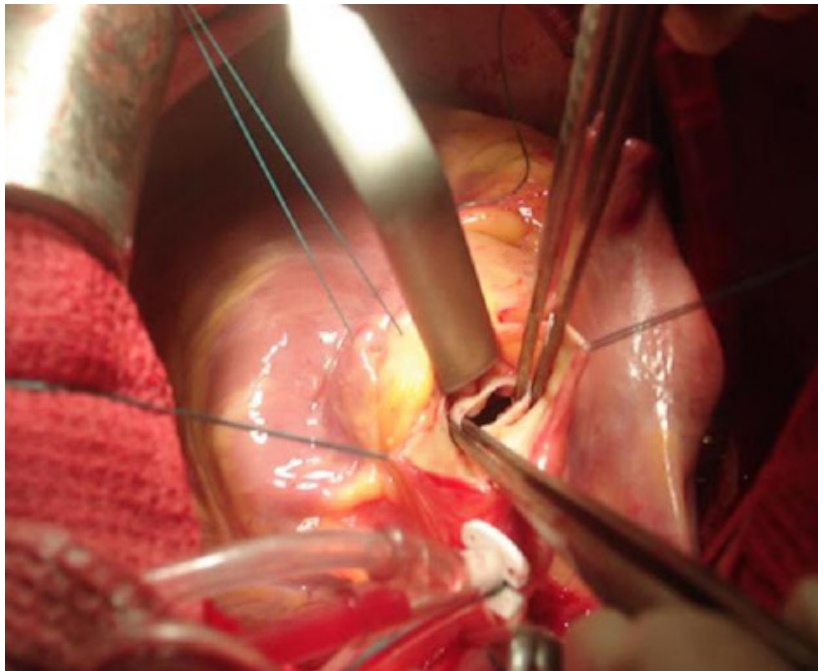


Figure 2. Bicuspid aortic valve during operation.

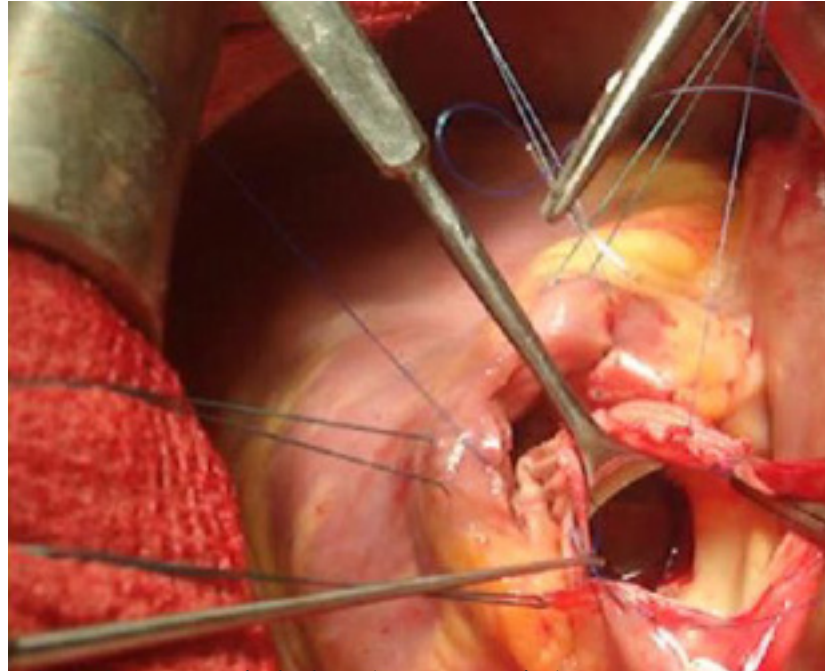


Figure 3. Aortoventriculoplasty (Konno operation) .

Discussion

Left ventricular outflow tract obstructions (LVOTOS) encompass a series of stenotic lesions starting in the anatomic left ventricular outflow tract (LVOT) and extending to the descending portion of the aortic arch. Obstruction may be subvalvar, valvar, and supra-valvar. These obstructions to forward flow may present alone or in concert, as in the frequent association of a bicuspid aortic valve with coarctation of the aorta. LVOTOS are congenital in the vast majority of individuals younger than 50 years old. The subaortic stenosis may be focal, as a discrete membrane, or more diffuse, resulting in a tunnel leading out of the left ventricle. The discrete form of fibro muscular subaortic stenosis is highly frequent, but tunnel type lesions are associated with a greater degree of stenosis. A bicuspid aortic valve is present in 23% of patients. All of these lesions impose

increased after load on the left ventricle and if severe and untreated, result in hypertrophy and eventual dilatation and failure of the left ventricle. It is imperative to consider all patients with LVOTOS at a high risk for developing infective endocarditis. It can be easily diagnosed by echocardiography and should be treated by early elective surgery. Surgical management consists of discrete membrane excision and / or blunt dissection in focal subaortic stenosis with focal septal myomectomy. Tunnel type subaortic stenosis is more surgically challenging and aortoventriculoplasty (Konno operation) is the reasonable procedure.⁷

Acknowledgements

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References

- 1 Marasini M, Zannini L, Ussia GP, et al. Discrete subaortic stenosis: incidence, morphology and surgical Impact of Associated subaortic Anomalies. *Ann thorac surg* 2003;**75**:1763-8.
- 2 Oliver JM, Gonzalez A, Gallego P, et al. Discrete Subaortic Stenosis in adults: increase prevalence and slow rate of progression of the obstruction and aortic regurgitation. *J Am Coll Cardiol* 2001;**38**:835-42. [11527642]
- 3 Brauner R, Laks H, Drinkwater D, et al. Benefits of early surgical repair in fixed subaortic stenosis. *J Am coll cardiol* 1997;**30**:1835-42. [9385915]
- 4 Konno S, Imai Y, Iida Y, et al. A new method for prosthetic valve replacement in congenital Aortic Stenosis associated with hypoplasia of Aortic valve ring. *J thorac cardio vasc surg* 1975;**70**:909-17. [127094]
- 5 Misbach GA, Turley K, Ullyot DJ, et al. Left ventricular out flow enlargement by the konno Procedure. *J Thorac Cardiovasc Surg.* 1982;**84**:696-703. [6215542]
- 6 Coskun KO, Bairaktoris A, Coskum ST, et al. Aortico –left ventricular Tunnel , an unusual congenital cardiac Anomaly in Adults, Application of a new operative technique. *ASAIO J.* 2006;**52**:e40-2. [17117046]
- 7 Rastan H, Koncz J. Aortoventriculoplasty: A new technique for the treatment of left ventricular out flow tract obstruction. *J Thorac Cardiovasc Surg* 1976;**71**:920-7.