Operative Techniques in Thoracic and Cardiovascular Surgery

Supramitral Obstruction of Left Ventricular Inflow Tract by Supramitral Ring

Igor Konstantinov, MD, Tae-Jin Yun, MD, Christopher Calderone, MD, and John G. Coles, MD

Supramitral ring (SMR) is a rare developmental abnormality of the supravalvular area of the mitral valve which produces a variable degree of obstruction to left ventricular filling. The morphological substrate of SMR consists of a fibro-membranous ring which is adherent to the atrial aspect of the mitral valve leaflets. SMR is morphologically and embryologically distinct from the more common entity of cor triatriatum, which consists of an obstructive fibro-muscular membrane located within the left atrium proximal to the left atrial appendage. Surgical resection of SMR is usually effective as evident by the generally benign postoperative hemodynamic outcome. Coexistent mitral valvular lesions are usual but not typically severe in degree.

KEYWORDS congenital mitral stenosis, surgical repair of supramitral ring, left heart inflow anomalies

Supramitral ring (SMR) is a rare developmental abnormality of the supravalvular area of the mitral valve (MV) producing obstruction to left ventricular filling.[1,2,3] SMR should be differentiated from cor triatriatum sinister, since the embryological origin, morphology and the surgical implications are different in these two malformations. Supramitral ridge is an extremely rare entity which may mimic classical SMR, but one anomaly which should be distinguished by the general rule that surgical resection is contraindicated. Although cases of isolated SMR have been reported sporadically.[5,6,7] SMR is more commonly associated with other obstructive lesions in the left heart such as valvular stenosis of MV, parachute MV or mitral arcade, subaortic membrane or fibromuscular tunnel, aortic valve stenosis with or without bicuspid aortic valve, transverse arch hypoplasia or discrete coarctation of aorta,[8,9] and less commonly with anomalies in the right heart such as pulmonary stenosis or tetralogy of Fallot.[10,11] Classically, SMR was described as one of the four specific features of Shones complex.[8] In his original description, Shone reported 8 patients who had multiple obstructive lesions in the left heart. Although only 2 patients exhibited all four features, SMR was a common finding in all 8 patients. Thus, SMR has been regarded as an essential element constituting Shone's complex. It is widely recognized, however, that only a small proportion of cases of SMR is associated with classical Shone's complex, as confirmed by a review of our own institutional experience.

Division of Cardiovascular Surgery, The Hospital for Sick Children, University of Toronto, Toronto, ON M5G 1X8, Canada.

Address reprints to Dr. John G. Coles, Division of Cardiovascular Surgery, Hospital for Sick Children, 555 University Avenue, Toronto ON M5G 1X8 Canada; E-mail: john.coles@sickkids.ca

The embryologic origin of supramitral ring is unclear, but it has been postulated that this condition results from incomplete division of endocardial cushion tissue.[7] Supramitral ring is differentiated from cor triatriatum, which is believed to be a result of incomplete absorption of primary pulmonary vein during the fifth embryonic week.[12,13] In cor triatriatum, the communication of proximal and distal chambers is usually a single hole in the membrane, which is further above the MV and proximal to left atrial auricle. Microscopically, SMR has a dense layer of sclerotic tissue resembling valve substance but without the discrete layers of a valve, while the membrane of cor triatriatum is characterized by bilaminar muscular structure.[14] SMR should also be differentiated from left ventricular inflow obstruction caused by dilated coronary sinus, and from the rare, isolated invagination of left atrial free wall immediately proximal to the mitral valve, referred to as supramitral ridge.

Safe surgical resection of the obstruction created by cor triatriatum sinister and SMR requires a thorough understanding of the anatomical details specific to each entity. The pathophysiology of all three anomalies is similar, however, and depends on the severity of the obstruction which the left atrium. In the case of mild obstruction, most blood will flow through the mitral valve (MV), creating the picture of mitral stenosis. Moderate to severe obstruction results in left to right shunting across the atrial septal defect (ASD), which is inversely proportional in volume to that of the antegrade flow across the left atrial obstructing lesion. The absence of an associated ASD, although unusual in all these entities, leads to the hemodynamic and clinical picture simulating total anomalous pulmonary venous drainage with obstruction.

1522-2942/04/$-see front matter © 2004 Elsevier Inc. All rights reserved.
doi:10.1053/j.optechstcvs.2004.07.001

Surgical Technique

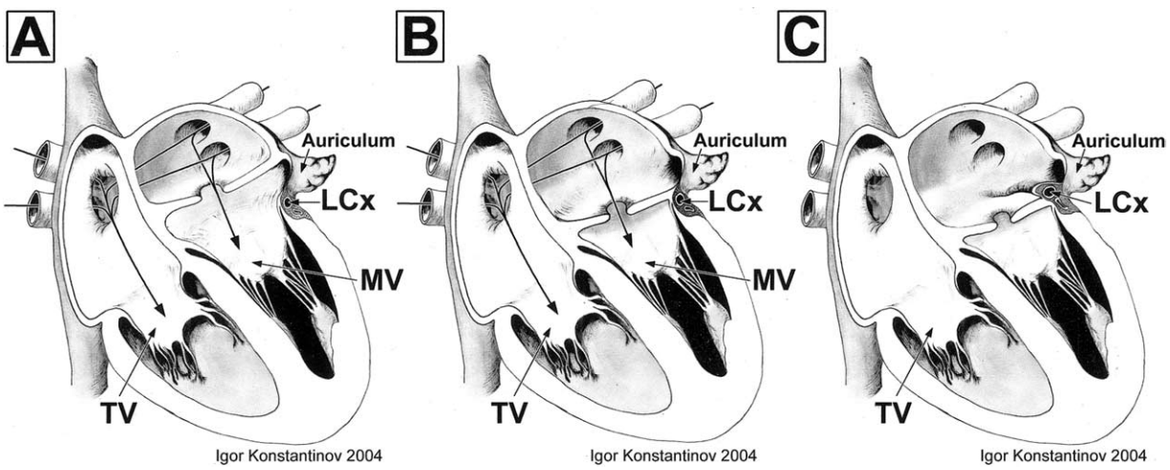

Figure 1 The morphology of each of these anomalies has distinct features of crucial importance to the surgeon. Cor triatriatum sinister (A) is a relatively common anomaly and the surgical removal of the intraatrial membrane is a comparatively straightforward operation detailed elsewhere.[15] In contrast SMR (B) is a very rare anomaly and resection of SMR is potentially more hazardous due to the intimate relationship of the fibro-membranous ring with the mitral valve and the close proximity of the left circumflex coronary artery (LCx). The location of the left atrial (LA) appendage (auriculum) may help to differentiate those two anomalies. In cor triatriatum sinister, the intraatrial membrane is located above the orifice of the left atrial appendage (A), whereas a SMR is located below it (B). Supramitral ridge, which must be extremely rare and is presented in the literature by a single case report, may simulate SMR but does not produce stenosis of the left atrial outflow, and thus does not require resection. Furthermore, resection of the supramitral ridge may damage the epicardial coronary vessel contained within the invaginated atrioventricular sulcus tissue.

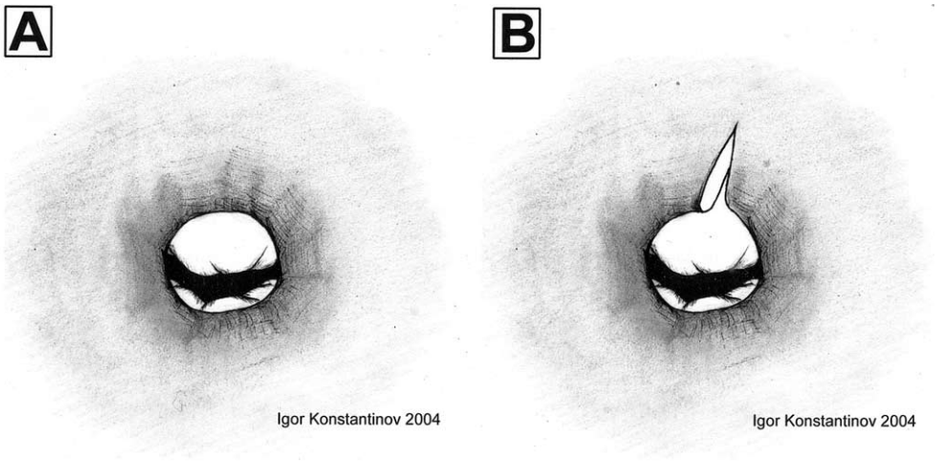

Figure 2 Our surgical approach to SMR consists of the following steps. After standard midline sternotomy, bicaval cannulation, initiation of cardiopulmonary bypass, and cardiac arrest, the LA is opened using a trans-septal approach. On entering the LA, the restrictive orifice of the membrane is identified (A). The location of the LA appendage orifice is determined and the mitral valve is carefully inspected through the orifice in the membrane. Stay sutures (not shown in the figure) are placed in the membrane, which are useful for applying traction and delineating the adherent membrane from the underlying valve leaflets. The initial incision is made anteriorly (above the anterior leaflet of the mitral valve and directed toward the aortic-mitral fibrous continuity) and extended to the junction of the membrane and the mitral valve annulus (B). This initial incision is directed away from the possible location of the LCx coronary artery. Care is taken to protect the mitral valve during the resection.

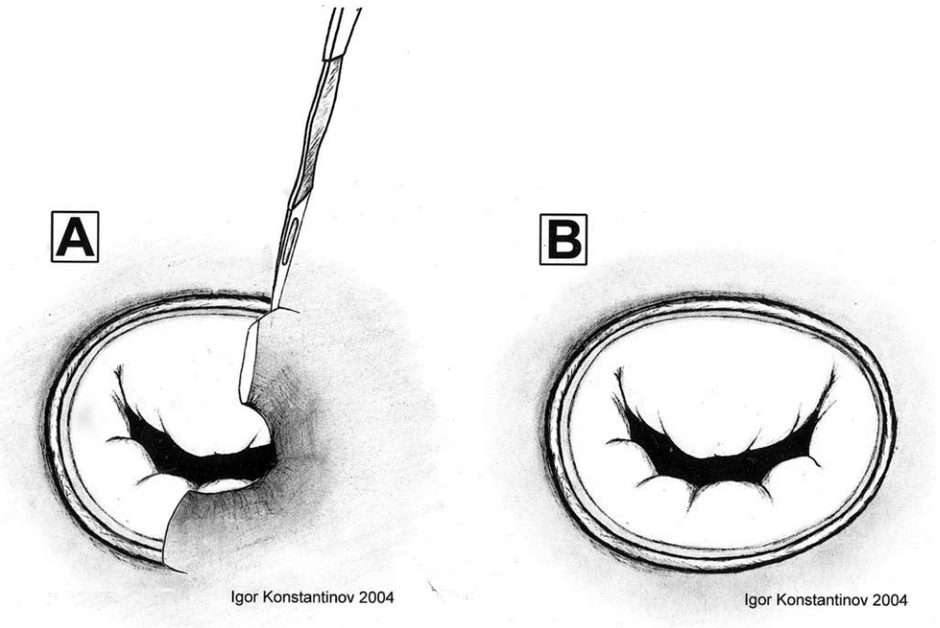

Figure 3 The anterolateral part of the membrane is removed first keeping in mind the proximity of the LCx coronary artery (A). The membrane is then resected completely (B). In our experience, mitral valve abnormalities, including variable degrees of annular hypoplasia, leaflet thickening and shortened chordae, often coexist with SMR. Specific surgical correction directed to these associated mitral valve anomalies is usually not effective, although papillary muscle splitting may improve leaflet excursion to some degree without creating incompetence. Nevertheless, the diastolic gradients across the mitral valve region are usually substantially reduced following SMR resection. Mitral valve replacement may be needed on rare occasions.[3]

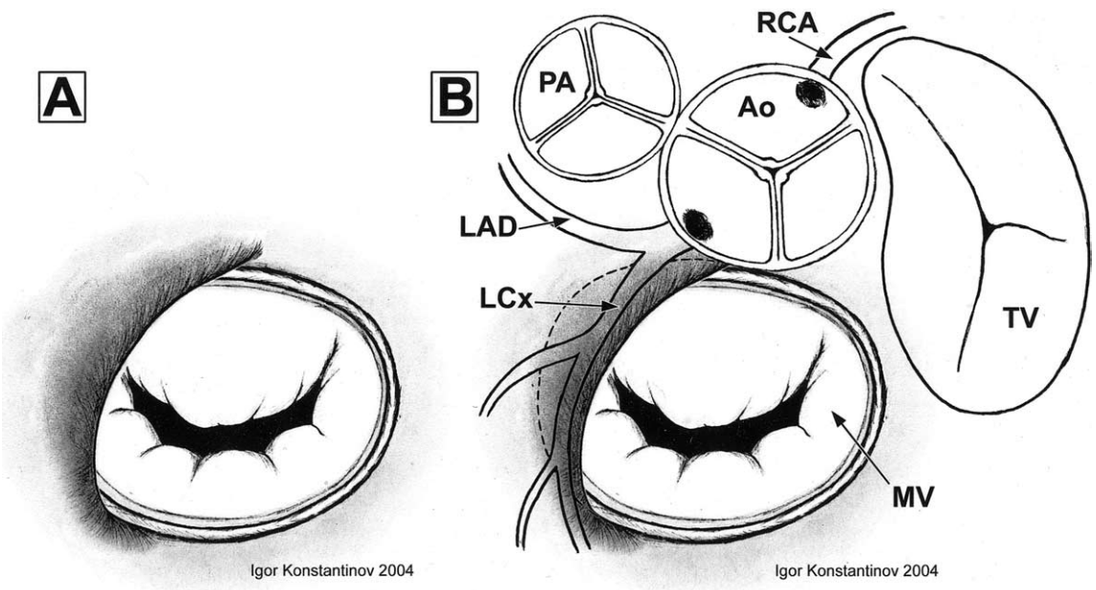

Figure 4 The finding of an eccentric, nonfibrous thickening in the area of the LCx coronary artery indicates the rare variant lesion, referred to as a supramitral ridge, which as indicated earlier should not be resected since it very likely harbors the coronary artery as shown in Fig. 4AB.[4]

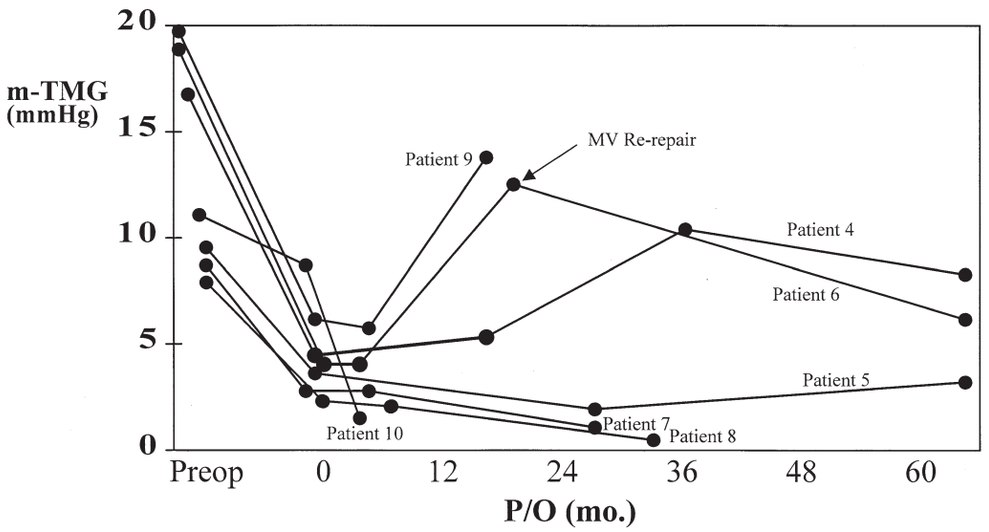

Figure 5 Postoperative changes in mean transmitral gradient (m-TMG).

Experience of the Hospital for Sick Children, Toronto

From 1983 to 2003, 13 patients were diagnosed with supravalvular obstruction of left ventricle inflow excluding cor triatriatum at the Hospital for Sick Children in Toronto. One patient, who had persistent left superior vena cava with dilated coronary sinus causing supramitral obstruction, was excluded from the analysis. Two additional patients, who were alleged to have SMR with mild stenosis at the time of their coarctation repair, were also excluded from this study since they have not received a surgical intervention for SMR. Thus, 10 patients with SMR underwent surgical treatment directed to the stenosing SMR.

Associated Anomalies

The MV annulus was designated as smallish either by preoperative echocardiography or based on inspection at the time of surgery in 7 of 10 patients, whereas left ventricle was deemed adequate to support biventricular repair in all, although one patient required a Norwood operation after a trial of biventricular repair. With respect to the morphology of MV, all except one were deemed to have MV abnormalities such as small MV annulus, valvular mitral stenosis, short chordae, or parachute MV. Two patients had concomitant aortic arch repair. The preoperative transmitral gradients ($n = 6$) ranged from 8 mm Hg to 17 mm Hg with a median value of 12.5 mm Hg.

Results

There were no early or late postoperative deaths. The membranous component of the SMR was completely excised in all but one case, in which delineation from the mitral valve proper was not successful. Generally, effective relief of supravalvular stenosis was confirmed by low mean transmitral gradient (3 mm Hg) determined by early postoperative echocardiography (Fig. 5). The median follow-up duration was 54.8 months (range 2 to 160 months). Two patients required reoperation for recurrent left ventricular inflow obstruction: one patient with classical Shone's complex with a prior coarctation repair underwent MV replacement (Carbomedics 16t mm) 29 months after SMR resection and required a redo MV replacement 62 months later (St Jude 23 mm); the other patient with isolated SMR required resection of recurrent SMR 26 months after the initial SMR resection. On follow-up echocardiography two additional patients had mean transmitral gradients greater than 5 mm Hg related to supravalvular obstruction.

Summary

SMR is an uncommon lesion which can be safely and effectively managed by surgical resection. Our relatively limited experience indicates that SMR is not usually associated with severe, multilevel left heart obstructive lesions or classical Shones syndrome. While coexistent mitral valvular lesions are usual, they are not typically severe in degree, as evident by the generally benign postoperative outcome following resection of SMR.

References

1. Coles JG, Williams WG, Watanabe T, et al: Surgical experience with reparative techniques in patients with congenital mitral valvular anomalies. Circulation 76:III117-III122, 1987
2. Mychaskiw G, Sachdev V, Braden DA, et al: SMR: An unusual cause of congenital mitral stenosis. Case series and review. J Cardiovasc Surg 43:199-202, 2002
3. Sullivan ID, Robinson PJ, de Leval M, et al: Membranous supravalvular mitral stenosis: A treatable form of congenital heart disease. J Am Coll Cardiol 8:159-164, 1986
4. Huhta JC, Edwards WD, Danielson GK: Supravalvular mitral ridge containing the dominant left circumflex coronary artery. J Thorac Cardiovasc Surg 81:577-579, 1981
5. Chung KJ, Manning JA, Lipchik EO, et al: Isolated supravalvular stenosing ring of left atrium; diagnosis before operation and successful surgical treatment. Chest 65:25-28, 1974
6. Sullivan ID, Robinson PJ, DeLeval M, et al: Membranous supravalvular mitral stenosis: A treatable form of congenital heart disease. J Am Coll Cardiol 8:159-164, 1986
7. Srinivasan V, Lewin AN, Pieroni D, et al: Supravalvular stenosing ring

of the left atrium: Case report and review of the literature. Cardiovasc Dis Bull Texas Heart Inst 7:149-158, 1980
8. Shone JD, Sellers RD, Anderson RC, et al: The developmental complex of "parachute mitral valve," supravalvular ring of the lest atrium, subaortic stenosis and coarctation of aorta. Am J Cardiol 11:714-725, 1963
9. Manganas C, LLiopoulos J, Chard RB, et al: Reoperation and coarctation of the aorta: The need for lifelong surveillance. Ann Thorac Surg 72(4):1222-1224, 2001
10. Benry J, Leachman RD, Cooley DA, et al: Supravalvular mitral stenosis associated with tetralogy of Fallot. Am J Cardiol 37:111-114, 1976
11. Horn AR, Jain KK, Tamer DM: Supravalvular mitral stenosis in a patient with tetralogy of Fallot. Am J Cardiol 22:733-737, 1968
12. Jacobstein MD, Hirschfield SS: Concealed left atrial membrane: pitfalls in the diagnosis of cor triatriatum and supravalve mitral ring. Am J Cardiol 49:780-786, 1982
13. Marin-Garcia J, Tandon R, Lucas RV, et al: Cor triatriatum: Study of 20 cases. Am J Cardiol 35:59-66, 1975
14. Oglietti J, Reul GJ, Leachman RD, et al: Supravalvular stenosing ring of the left atrium. Ann Thorac Surg 21:421-424, 1976
15. Dipchand Ai, Freedom RM, Yoo S-J: The divided left atrium (Cor Triatriatum). The Natural and Modified History of Congenital Heart Disease. Toronto, Canada, Blackwell Publishing 24C:295-298, 2004