Published in final edited form as:

J Am Soc Echocardiogr. 2015 May ; 28(5): 541–548. doi:10.1016/j.echo.2015.01.013.

Echocardiographic Characteristics of Annulo-Leaflet Mitral Ring

David N. Schidlow, MD, MMus¹, Abbas Zaidi, MD², Kimberlee Gauvreau, ScD¹, Sitaram M. Emani, MD³, Tal Geva, MD¹

¹Department of Cardiology, Boston Children's Hospital

²Department of Pathology, Boston Children's Hospital

³Department of Cardiac Surgery, Boston Children's Hospital

¹Department of Pediatrics, Harvard Medical School, Boston, MA

²Department of Pathology, Harvard Medical School, Boston, MA

³Department of Surgery, Harvard Medical School, Boston, MA

Abstract

Background—Annulo-leaflet mitral ring (ALMR) is a rare congenital cardiac anomaly caused by fibrous tissue on the atrial surface of the mitral valve, which can progress to cause severe stenosis. Because little information is available on the detailed echocardiographic features of the anomaly and their associations with surgical resection, this study was undertaken to address these questions.

Methods—Retrospective single-center study of clinical and echocardiographic data of patients with ALMR from 2004 through 2012. Data were analyzed for association with surgical resection.

Results—The median age at diagnosis of the 57 study patients was 1.8 years (63% males). Isolated ALMR was found in 6 patients (11%). The remaining 51 (89%) patients had associated lesions: additional mitral valve abnormalities in 35 (61%), coarctation in 11 (19%), and subaortic stenosis in 15 (26%). ALMR was best visualized in the apical 4-chamber view, and the lesion was indistinct from the parasternal long-axis view in 25% of patients. We identified 7 patterns of ALMR, differentiated by leaflet involvement (anterior, posterior, or both) and location (annular vs. intraleaflet). Compared with other patterns, intraleaflet morphology had a higher mean MS gradient than those without (8.4 vs. 5.8 mm Hg, $p=0.01$). Among the 57 patients, 32 (56%) underwent ALMR resection at a median age of 1.5 years. Younger age at echocardiographic diagnosis ($p=0.02$) and short chordae ($p=0.03$) were associated with resection.

Address for correspondence: David N. Schidlow, MD, MMus, Children's National Heart Institute, Children's National Medical Center, 111 Michigan Avenue Northwest, Washington, DC 20010, Business Tel: (202) 476-2020, Fax: (202) 476-5700, dschidlo@childrensnational.org.

Conflicts of interest:

None

Publisher's Disclaimer: This is a PDF file of an unedited manuscript that has been accepted for publication. As a service to our customers we are providing this early version of the manuscript. The manuscript will undergo copyediting, typesetting, and review of the resulting proof before it is published in its final citable form. Please note that during the production process errors may be discovered which could affect the content, and all legal disclaimers that apply to the journal pertain.

Conclusions—The morphology of ALMR as evaluated by echocardiography is heterogeneous and can be classified based on lesion location and leaflet involvement. Intraleaflet morphology is associated with significant mitral stenosis; younger age at diagnosis and short chordae are associated with ALMR resection.

Annulo-leaflet mitral ring (ALMR) is a rare form of mitral stenosis and often presents a diagnostic challenge. This lesion has multiple pseudonyms, including “supramitral” ring, despite several investigations suggesting that the lesion is not truly supra-annular.^{1–3} ALMR has been defined as a ridge of connective tissue adherent to the atrial surface of the mitral valve or slightly superior to the annulus and distal to the left atrial appendage.^{4,5} First recognized in 1902, ALMR is often associated with additional left-sided obstructive lesions, including mitral valve, aortic valve, and aortic arch abnormalities.^{6,7} Shone and colleagues described ALMR in 1963 as part of a complex association of left heart obstructive lesions, which included parachute mitral valve, discrete subaortic stenosis, and coarctation of the aorta. The ALMR, called “supravalvar ring of left atrium” in their study, was noted to be “present but not functionally significant” in 5 of the 8 studied patients, an observation that highlights the substantial heterogeneity of this lesion.⁸ In addition to its association with left heart obstructive lesions, ALMR has also been described in the setting of a variety of other congenital cardiac anomalies, including transposition of the great arteries and tetralogy of Fallot.^{9–13}

ALMR was first demonstrated by M-mode echocardiography in 1972 and later by 2-dimensional echocardiography in 1980. These early studies demonstrated that ALMR presented a diagnostic challenge.^{14–17} Since these initial descriptions of ALMR, several studies have described pathologic, histologic, and surgical aspects of the anomaly.^{4,16,18,19} Surgical reports note that when isolated, the lesion is amenable to resection with good outcome,¹⁹ but results are often less satisfactory when ALMR is associated with other lesions.^{1,20–22} Balloon valvuloplasty is recognized as having limited utility in the setting of ALMR.^{23,24}

A paucity of information is available regarding echocardiographic characteristics of ALMR. Toscano et al., in the largest study to date (n=25), importantly recognized 2 different variants of this anomaly, namely “supramitral” and “intramitral”.¹ They noted that supramitral lesions were less likely to have associated abnormalities and have better prognosis, as compared with intramitral lesions. However, detailed characterization of the lesion by echocardiography is lacking. Therefore, we undertook this study to (1) characterize the echocardiographic features of ALMR in a relatively large cohort; (2) create a classification system based on lesion location; and (3) identify echocardiographic characteristics associated with resection.

METHODS

Patients

The database of the Boston Children’s Heart Center was searched for patients with the diagnostic code of supravalvar mitral stenosis. Included patients had echocardiograms from March 2004 through December 2012. This period was selected to coincide with the

utilization of digital image archiving. Exclusion criteria included the following: (1) prior surgical or catheter-based intervention on the mitral valve; (2) diagnosis of atrioventricular canal defect; and (3) patients with functional single ventricle. Medical records were reviewed for demographic and clinical data. Demographic data, presence of genetic syndromes, and a history of prior surgical or catheter-based interventions were recorded. Additional clinical data included outpatient notes, catheterization reports, and surgical reports. Primary echocardiographic data was reviewed as described below. The Boston Children's Hospital Committee on Clinical Investigations approved the study protocol and waived the requirement for informed consent.

Outcome

The primary clinical outcome was surgical resection of ALMR.

Echocardiography

Echocardiograms were performed using either a Hewlett Packard 7500 or a Philips iE33 scanner. Digitally archived studies were reviewed using HeartSuite VERICIS workstations (Emageon, Hartland, WI, USA). Studies were analyzed for left atrium, left ventricle, mitral valve, aortic valve, and aortic arch dimensions and morphology. A single reviewer (DNS) who was blinded to patient outcomes performed all of the measurements listed below. Categorization of ALMRs was made by consensus of 2 investigators (DNS and TG) who were blinded to patient outcomes based on pre-defined criteria.

For patients with serial echocardiograms, the preoperative study was utilized for data collection. For patients who did not have surgery, the highest-quality study at diagnosis was used. Patients with serial echocardiograms had the mitral valve gradient recorded at each study.

The following data were collected: (1) mitral valve diameters (anteroposterior and lateral) and Z-scores, (2) left ventricular end-diastolic volume and Z-score, (3) left ventricular mass and Z-score, (4) ejection fraction, (5) left atrial (LA) area adjusted to body surface area, and (6) mean mitral valve gradient. Body surface area was calculated from the patient height and weight using the Haycock formula.²² Z-scores were calculated using our laboratory's published normative data.²³

Echocardiograms were reviewed for the following associated abnormalities: (1) short chordae, (2) closely spaced papillary muscles, (3) parachute mitral valve, (4) subaortic stenosis, (5) aortic valve morphology (categorized as tricommissural, bicommissural, or unicommissural), and (6) additional congenital cardiac anomalies. Shortened chordae and closely spaced papillary muscles were determined based on qualitative assessment. The designation of shortened chordae was assigned when there was markedly diminished chordal length and a short distance from the edge of the mitral leaflets to the papillary muscles on the apical 4-chamber view. The designation of closely spaced papillary muscles was assigned when the normal interpapillary distance was markedly reduced when assessed by the parasternal or subcostal short-axis views in diastole. For patients with serial echocardiograms, the initial mitral valve gradient at the time of diagnosis was recorded. The following measurements of the ALMR were performed from the apical 4-chamber view in

diastole: (1) membrane length, (2) mitral valve leaflet lengths, and (3) location of obstructive ridge (categorized as annular or intraleaflet). Annular lesion was defined as a tissue ridge extending from the atrioventricular junction toward the lumen without significant adherence to the atrial surface of the leaflet tissue. An intraleaflet lesion was defined as a tissue ridge extending from the atrioventricular junction along the atrial surface of one or both mitral leaflets with the leading edge protruding into the lumen some distance from the annulus.

To allow analysis of the degree of lesion extension onto the mitral leaflets as a continuous variable, we measured the length of the obstructive membrane from the atrioventricular junction to its edge as well as the length of the respective leaflet. These measurements were then used to calculate the extent of the lesion, termed the membrane extent index (MEI), using the following formula: $[(\text{anterior membrane length} \div \text{anterior leaflet length}) + (\text{posterior membrane length} \div \text{posterior leaflet length})]/2$ (Figure 1). Measurements were performed exclusively in the apical 4-chamber view during diastole. Care was taken to exclude the chordae tendineae from measurements of the mitral leaflet length based on review of cine loops. As indicated by the formula, anterior and posterior indices were averaged to yield the MEI.

Statistical Analysis

Continuous variables were summarized using the median and range, unless otherwise noted. Categorical variables were summarized as frequencies and percentages and were compared using Fisher's exact test. Comparison was made between patients who had ALMR resection and those who did not using the Wilcoxon rank-sum test or the unpaired t-test for continuous variables, as appropriate. Logistic regression was used to estimate odds ratios and 95% confidence intervals of ALMR resection. A P value <0.05 was considered statistically significant. Statistical analysis was performed using commercially available statistical analysis software (Stata version 12, College Station, Texas).

RESULTS

Patients

Demographic, clinical, and prior surgical characteristics of the 57 patients meeting inclusion criteria are summarized in Table 1. The majority of patients were diagnosed before age 2 years, but the age at diagnosis ranged from 0 to 76.9 years. Of the 57 patients, 32 (56%) had prior non-mitral valve surgery. This most commonly included coarctation repair in 23 patients (40%) followed by ventricular septal defect closure in 5 (9%). Genetic abnormalities were present in 11 patients (19%), but there was no predominant syndrome among this group. Diagnostic or interventional catheterization was performed in 22 patients (39%). Of the 32 patients who underwent ALMR resection, 17 (53%) underwent pre-operative catheterization (16 diagnostic and 1 balloon mitral valvuloplasty). In contrast, among the 25 patients who did not undergo ALMR resection, 5 (20%) underwent catheterization (4 diagnostic and 1 balloon mitral valvuloplasty).

Echocardiographic Data

ALMR and Associated Anomalies—Echocardiographic data pertaining to the mitral valve, left ventricle, aortic valve, and aortic arch are summarized in Table 2. ALMR was not associated with other cardiovascular anomalies (i.e., “isolated”) in only 6 of the 57 patients (11%). Associated mitral valve abnormalities were common and included short chordae in 23 patients (40%), closely spaced papillary muscles in 19 (33%), and parachute deformity in 8 (14%). Mitral regurgitation was present in 14 patients (25%), and was mild in 12. Subvalvar and valvar aortic stenoses were noted in 25 and 26% of patients, respectively. The aortic valve was frequently abnormal with bicommissural or unicommissural valve in 29 patients (51%), whereas coarctation of the aorta was diagnosed in 11 (19%). ALMR was part of a Shone association in 5 patients (9%).

Imaging Considerations—ALMR was best visualized in the apical 4-chamber plane. Prominent lesions were also seen in the parasternal long-axis views but were often less distinct. Importantly, in 25% of patients the lesion was indistinct when imaged from the parasternal long-axis view and the diagnosis was made exclusively from the apical window. Side-by-side display of two-dimensional and color-coded Doppler images was particularly helpful with the latter showing the location of flow acceleration while the former depicting the anatomic details. Three-dimensional imaging was available only in a small number of cases and its contribution to the diagnostic evaluation could not be assessed in this study.

Classification of ALMR Morphology—Types of ALMR were categorized based on leaflet involvement (anterior, posterior, or both) and whether the lesion was protruding from the basal portion of the leaflets into the lumen (annular ALMR) or adherent and extending onto the atrial surface of the leaflet (intraleaflet ALMR). Based on these criteria, we identified 7 distinct types of ALMR, which are shown diagrammatically in Figure 2 and by echocardiography in Figure 3. As expected, the MEI was significantly higher in patients with intraleaflet involvement as compared with annular lesions (0.5 ± 0.21 vs. 0.27 ± 0.27 , $p < 0.001$). Moreover, compared with the annular type, the mean mitral valve gradient was significantly higher in the intraleaflet type (8.4 ± 3.7 vs. 5.8 ± 3 mm Hg, $p = 0.01$).

Factors Associated with ALMR Resection—Of the 57 patients, 32 (56%) underwent ALMR resection at a median age of 1.5 years (range 0 – 16.8 years). Compared with patients who did not undergo ALMR resection, those who did tended to be younger at the time of diagnosis (median age 1.6 vs. 3.7 years, $p = 0.24$), had a significantly lower body surface area (median 0.42 vs. 0.8 m², $p = 0.01$), a larger left atrial area index (17.5 vs. 9.4 cm²/m², $p = 0.005$), and more frequently had short chordae (53% vs. 24%, $p = 0.03$). In addition, the mean mitral inflow Doppler gradient at the first echocardiogram was higher in those who underwent resection (median 7.5 mm Hg vs. 5 mm Hg, $p = 0.06$) with a subsequent increase in the gradient in the resection group as oppose to non-progression in those who did not (latest mean gradient 9 mm Hg vs. 5 mm Hg, $p < 0.001$). Logistic regression analysis identified younger age at diagnosis and short chordae as factors associated with resection (Table 3). A larger left atrial area index tended to be related to ALMR resection ($p = 0.06$), but this did not reach statistical significance. The morphologic

type of ALMR was not significantly different between those who did or did not undergo resection.

Surgical Interventions—Surgical resection of the ALMR was performed in 32 (56%) patients at a median interval of 4 months after diagnosis (Table 4). Indications for surgery were often based on a combination of anatomic, hemodynamic, and clinical findings and varied considerably among patients and providers. Of the 32 patients who underwent ALMR resection, 18 had additional surgical procedures performed on the mitral valve, including mobilization and splitting of papillary muscles in 9, enlargement of interchordal spaces in 6, leaflet augmentation in 5, and commissuroplasty in 4. Concomitant non-mitral surgery (n=24) included resection of discrete subaortic stenosis in 10, ventricular septal defect closure in 9, coarctation of the aorta repair in 5, aortic valvuloplasty in 3, endocardial fibroelastosis resection in 2, aortic valve replacement in 1, and tetralogy of Fallot repair in 1. In addition to the 32 patients who underwent ALMR resection, 6 patients with mild ALMR underwent non-mitral valve surgical procedures.

All patients survived ALMR surgery and associated procedures. Early results of ALMR resection were good in all 32 patients with a decrease in the mean transmitral gradient from a median of 9 mm Hg to 2 mm Hg ($p < 0.001$). The median duration of follow-up was 1.5 years (range 0.1 – 17.5 years). During this period, the mean mitral valve gradient remained stable or decreased slightly in 28 patients. However, 4 patients (13%) had recurrence of ALMR, and 2 of these underwent repeat resection 5 months and 2 years after the first operation, respectively.

DISCUSSION

In this largest cohort of patients with ALMR studied to date, we propose an echocardiography-based classification scheme for characterizing the morphologic features of the anomaly based on lesion location and leaflet involvement. Together with demographic and other anatomic features of the mitral apparatus, this classification aids in the assessment of disease progression and identifies echocardiographic characteristics associated with resection.

The classification of ALMR morphology described in this paper first distinguishes between annular and intraleaflet types. In both types, the obstructive tissue involves the fibrous junction between the left atrium and left ventricle, the mitral valve annulus. In the annular type, the fibroelastic tissue extends from the annulus (atrioventricular junction) onto the lumen with little or no attachment to the leaflet tissue. In the intraleaflet type, the fibroelastic tissue extends from the annulus onto the atrial surface of the leaflet with a free edge protruding into the lumen deep into the involved leaflet. Once the distinction between annular and intraleaflet type is made, the involvement of the anterior and posterior leaflets is described. In this cohort, an intraleaflet lesion involving either one or both leaflets was associated with more severe mitral stenosis as evident by a higher mean mitral gradient by Doppler.

The classification scheme described in this paper expands on the observations of Toscano et al. who noted that ALMR could be divided into supramitral (our annular) and intramitral (our intraleaflet) categories.¹ Similar to our findings, their intramitral lesions were more often associated with progressive mitral stenosis. However, in addition to the detailed classification provided in this report, we note that none of our cases had a truly supramitral lesion location, defined as obstructive fibroelastic tissue located within the left atrium above (upstream) to the fibrous atrioventricular junction (annulus). Given that a truly supramitral location has either not been documented or is exceedingly rare, we propose that the term *supramitral ring* be replaced by the more precise term *annulo-leaflet mitral ring*, or ALMR.

Our proposed revised nomenclature addresses an important deficiency of the current often used term “supramitral ring”. The term “annulo-leaflet” correctly indicates that the lesion arises at the annular level (the atrioventricular junction) and emphasizes the intimate involvement of the fibroelastic tissue with the mitral valve leaflets. No patients in our series had a truly supra-annular membrane, and renaming this lesion will help alleviate any misconceptions that the lesion should be in that position. This revised term may direct the attention of sonographers and those interpreting echocardiograms to the atrioventricular junction and mitral valve leaflets rather than the left atrium. In addition, the classification scheme of lesion location and mitral extent index described in this study augment the revised nomenclature by acknowledging the marked heterogeneity of ALMR.

It is interesting to note that several reports have described a stenosing ring associated with the tricuspid valve in the setting of atrioventricular discordance.^{25–27} In this exceedingly rare anomaly, the obstructive fibrous tissue extends from the atrioventricular junction between the left atrium and right ventricle, involving the tricuspid valve. These cases highlight the observation that this lesion is not unique to the mitral valve. From the perspective of nomenclature, this anomaly can be more broadly described as “*annulo-leaflet atrioventricular valve ring*”; however, as the overwhelming majority of these cases occur in the setting of atrioventricular concordance and on the mitral valve, the term ALMR is appropriate. Nonetheless, we recognize that annulo-leaflet tricuspid ring (ALTR) exists.

As noted by previous authors, the echocardiographic diagnosis of ALMR is challenging with several reports noting missed diagnoses.^{1,2,17,28} The importance of accurate diagnosis is highlighted by the observations that unlike other types of congenital mitral stenosis, ALMR is amenable to surgical resection and does not respond favorably to transcatheter therapy.²⁴ Recognition by echocardiography requires a high index of suspicion, and should be considered in every patient with congenital mitral stenosis. The ALMRs in our cohort were most readily visualized from the apical 4-chamber view as compared with the parasternal long-axis view. This is likely due to signal dropout resulting from the nearly parallel angle of insonation of the ultrasound beam relative to the orientation of the obstructive tissue from the parasternal window. Other acoustic windows were seldom helpful. In many cases, the diagnosis was facilitated by the use of color Doppler, which showed increased flow velocity beginning at the level of the mitral valve annulus. We also noted that side-by-side display of the anatomy and color Doppler (Figure 4) was particularly helpful because it facilitates correlation between morphology and flow acceleration. Finally, although not common in our cohort, flow acceleration by color Doppler may not be apparent in patients with a significant

atrial septal defect or in patients with mildly obstructive lesions. The latter is particularly common when the lesion is limited to the posterior leaflet, as previously noted Collison et al.²

We identified several factors to be associated with resection of ALMR, including short chordae tendineae, younger age at diagnosis, and a higher indexed left atrial area. It is reasonable to speculate that younger age at diagnosis and left atrial enlargement are surrogates of a more aggressive disease process with higher left atrial pressures. The association between short chordae and surgical resection of ALMR suggests that patients with progressive disease often have mitral pathology that involves the subvalvar apparatus, including the chordae tendineae and papillary muscles. This is supported by the observation that of the 32 patients who underwent surgical resection of ALMR, 14 (44%) required additional mitral valve surgery. As noted by other investigators,^{1,20} surgical resection of ALMR is an effective treatment with short-term elimination or marked reduction of the mitral valve gradient in all surgical patients in this series. Recurrent ALMR after initial successful resection has been noted in smaller series,^{24,29} and was found in 4 (13%) cases in our series. However, the modest sample size and relatively short duration of follow-up hamper our ability to assess the natural history of the lesion and to identify patients at risk for recurrence. Interestingly, the rate of recurrence noted in our cohort is similar to published reports of recurrence of subaortic stenosis (14%).³⁰ Taken together with similarities in histopathology between these obstructive left heart obstructive lesions,³¹ it is tempting to speculate about possible shared origins of this disease.

Limitations

The retrospective nature of this investigation poses several limitations. Echocardiographic equipment and technology had evolved during the study period with earlier studies performed on older equipment with less refined image quality. However, we selected a time period during which studies were digitally archived, thus avoiding videotaped studies with lower quality images. We used LA area as a marker of LA size, instead of LA volume, because calculation of biplane LA volume was not possible in early cases due to lack of 2-chamber apical images. Another limitation related to the retrospective study design is lack of clinical and echocardiographic data from the newborn and early infancy periods in some patients who were referred to us for management of mitral stenosis from other institutions. Similarly, follow-up data are limited in patients who returned to their home institution. However, we made every effort to collect follow-up data whenever feasible.

Conclusions

ALMR is a rare congenital anomaly that seldom occurs in isolation. The morphology of the anomaly as evaluated by echocardiography is heterogeneous and can be classified based on lesion location and leaflet involvement. Intraleaflet morphology is associated with significant mitral stenosis, whereas younger age at diagnosis and short chordae are associated with progressive disease leading to ALMR resection.

Acknowledgments

Dr. Schidlow is supported by the National Institutes of Health (T32-HL007572). This study was supported in part by the Higgins Family Noninvasive Research Fund.

References

1. Toscano A, Pasquini L, Lacobelli R, Di Donato RM, Raimondi F, Carotti A, et al. Congenital supralvalvar mitral ring: an underestimated anomaly. *J Thorac Cardiovasc Surg.* 2009; 137:538–42. [PubMed: 19258061]
2. Collison SP, Kaushal SK, Dagar KS, Iyer PU, Girotra S, Radhakrishnan S, et al. Supramitral ring: good prognosis in a subset of patients with congenital mitral stenosis. *Ann Thorac Surg.* 2006; 81:997–1001. [PubMed: 16488709]
3. Vaideeswar P, Baldi MM, Warghade S. An analysis of 24 autopsied cases with supramitral rings. *Cardiol Young.* 2009; 19:70–5. [PubMed: 19079810]
4. Ruckman RN, Van Praagh R. Anatomic types of congenital mitral stenosis: report of 49 autopsy cases with consideration of diagnosis and surgical implications. *Am J Cardiol.* 1978; 42:592–601. [PubMed: 696643]
5. Cassano GB. Congenital Annular Stenosis of the Left Atrioventricular Canal. So-Called Supralvalvar Mitral Stenosis. *Am J Cardiol.* 1964; 13:708–13. [PubMed: 14152015]
6. Fisher T. Two Cases of Congenital Disease of the Left Side of the Heart. *Br Med J.* 1902; 1:639–41. [PubMed: 20760111]
7. Gilbert G, Aerichide N, Bourassa M, David P. Supralvalvar mitral stenosis associated with ventricular septal defect, subvalvular aortic stenosis and double aortic arch. *Am J Cardiol.* 1966; 18:605–9. [PubMed: 4224285]
8. Shone JD, Sellers RD, Anderson RC, Adams P Jr, Lillehei CW, Edwards JE. The developmental complex of “parachute mitral valve,” supralvalvar ring of left atrium, subaortic stenosis, and coarctation of aorta. *Am J Cardiol.* 1963; 11:714–25. [PubMed: 13988650]
9. Iyer AP, Haridas A, Pathak SA, Duara R, YP, Pradhan S, et al. Supracardiac total anomalous pulmonary venous connection with a supramitral ring: a rare, surgically correctable anomaly. *Ann Thorac Surg.* 2006; 82:322–3. [PubMed: 16798243]
10. Hohn AR, Jain KK, Tamer DM. Supralvalvar mitral stenosis in a patient with tetralogy of Fallot. *Am J Cardiol.* 1968; 22:733–7. [PubMed: 5683430]
11. Chesler E, Beck W, Barnard CN, Schrire V. Supralvalvar stenosing ring of the left atrium associated with corrected transposition of the great vessels. *Am J Cardiol.* 1973; 31:84–8. [PubMed: 4682415]
12. Benry J, Leachman RD, Cooley DA, Klima T, Lufschanowski R. Supralvalvar mitral stenosis associated with tetralogy of Fallot. *Am J Cardiol.* 1976; 37:111–4. [PubMed: 1244728]
13. Baysan O, Uzun M, Bulakbasi N, Erinc K, Genc C, Isik E. Univentricular heart and supramitral ring: an unusual association. *Int J Cardiovasc Imaging.* 2005; 21:645–7. [PubMed: 16322925]
14. Lundstrom N-R. Ultrasoundcardiographic Studies of the Mitral Valve Region in Young Infants with Mitral Atresia, Mitral Stenosis, Hypoplasia of the Left Ventricle, and Cor Triatriatum. *Circulation.* 1972; 45:324–34. [PubMed: 5009477]
15. Lundstrom N-R. Echocardiography in the Diagnosis of Congenital Mitral Stenosis and in Evaluation of the Results of Mitral Valvotomy. *Circulation.* 1972; 46:44–54. [PubMed: 5039823]
16. Chung KJ, Manning JA, Lipchik EO, Gramiak R, Mahoney EB. Isolated supralvalvar stenosing ring of left atrium: diagnosis before operation and successful surgical treatment. *Chest.* 1974; 65:25–8. [PubMed: 4271992]
17. Snider AR, Roge CL, Schiller NB, Silverman NH. Congenital left ventricular inflow obstruction evaluated by two-dimensional echocardiography. *Circulation.* 1980; 61:848–55. [PubMed: 7357725]
18. Rogers HM, Waldron BR, Murphey DF, Edwards JE. Supralvalvar stenosing ring of left atrium in association with endocardial sclerosis (endocardial fibroelastosis) and mitral insufficiency. *Am Hear J.* 1955; 50:777–81.

19. Uva MS, Galletti L, Gayet FL, Piot D, Serraf A, Bruniaux J, et al. Surgery for congenital mitral valve disease in the first year of life. *J Thorac Cardiovasc Surg.* 1995; 109:164–6. [PubMed: 7815793]
20. Brown JW, Ruzmetov M, Rodefeld MD, Turrentine MW. Surgical strategies and outcomes in patients with supra-annular mitral ring: a single-institution experience. *Eur J Cardiothorac Surg.* 2010; 38:556–60. [PubMed: 20620074]
21. Coles JG, Williams WG, Watanabe T, Duncan KF, Sherret H, Dasmahapatra HK, et al. Surgical experience with reparative techniques in patients with congenital mitral valvular anomalies. *Circulation.* 1987; 76:117–22.
22. Malhotra SP, Lacour-Gayet F, Campbell DN, Miyamoto S, Clarke DR, Dines ML, et al. Outcomes of reparative and transplantation strategies for multilevel left heart obstructions with mitral stenosis. *Ann Thorac Surg.* 2008; 86:1305–9. [PubMed: 18805182]
23. Spevak PJ, Bass JL, Ben-Shachar G, Hesslein P, Keane JF, Perry S, et al. Balloon angioplasty for congenital mitral stenosis. *Am J Cardiol.* 1990; 66:472–6. [PubMed: 2143623]
24. McElhinney DB, Sherwood MC, Keane JF, del Nido PJ, Almond CS, Lock JE. Current management of severe congenital mitral stenosis: outcomes of transcatheter and surgical therapy in 108 infants and children. *Circulation.* 2005; 112:707–14. [PubMed: 16043648]
25. Allwork SP, Phil M, Bentall HH, Cameron H. Congenitally Corrected Transposition of the Great Arteries: Morphologic Study of 32 Cases. *Am J Cardiol.* 1970; 38:910–23.
26. Quero-Jimenez M, Raposo-Sonnenfeld I. Isolated ventricular inversion with situs solitus. *Heart.* 1975 Mar 1; 37:293–304.
27. Kumar S, Phadke M, Kerkar P. Supratricuspid obstructive membrane in congenitally corrected transposition of the great arteries. *Ann Pediatr Cardiol.* 2013; 6:204–205. [PubMed: 24688250]
28. Piot JD, Rey C, Guerinon J, Lucet P, Pernot C, Binet JP. 2-dimensional echocardiography of congenital obstruction of left ventricle inflow. Apropos of 25 cases. *Arch Mal Coeur Vaiss.* 1984; 77:510–6. [PubMed: 6428347]
29. Tulloh RM, Bull C, Elliott MJ, Sullivan ID. Supravalvar mitral stenosis: risk factors for recurrence or death after resection. *Br Hear J.* 1995; 73:164–8.
30. Geva A, McMahon CJ, Gauvreau K, Mohammed L, del Nido PJ, Geva T. Risk factors for reoperation after repair of discrete subaortic stenosis in children. *J Am Coll Cardiol.* 2007 Oct. 50:1498–504. [PubMed: 17919571]
31. Ferrans VJ, Muna WF, Jones M, Roberts WC. Ultrastructure of the fibrous ring in patient with discrete subaortic stenosis. *Lab Invest.* 1978; 39:30–40. [PubMed: 682590]

Figure 1. Membrane extent index (MEI) measurements. The MEI is an average of anterior and posterior ALMR length compared to total leaflet length.

Figure 2.
Diagram showing anatomic types of ALMR.

Figure 3.
Echocardiographic examples of ALMR types.

Figure 4.

Echocardiographic imaging of ALMR from the apical 4-chamber view with side-by-side display of 2-dimensional and color Doppler. The ALMR extends from the annulus onto the lumen with proximal attachments to the leaflet tissue (arrows). Color comparison (right panel) demonstrates flow acceleration beginning at the level of the obstructive tissue (arrows). LA, left atrium; LV left ventricle; arrows, ALMR.

Table 1

Demographic and clinical variables

	All Patients (n=57)	ALMR Resection (n=32)	No ALMR Resection (n=25)	P-value
Age at diagnosis (y)	1.8 (0 – 76.9)	1.6 (0 – 16.8)	3.7 (0.1 – 76.9)	0.24
Age at echocardiogram (y)	2.1 (0 – 76.9)	1.7 (0 – 16.8)	4.2 (0.1 – 76.9)	0.08
BSA	0.49 (0.21 – 2.27)	0.42 (0.21 – 1.48)	0.80 (0.23 – 2.27)	0.01
Male	35 (61%)	19 (59%)	16 (64%)	0.79
Genetic syndrome *	11 (19%)	6 (19%)	5 (20%)	1.0
Associated anomalies	51 (89%)	30 (52%)	21 (37%)	0.89
Subaortic stenosis	15 (26%)	11 (34%)	4 (16%)	0.14
Aortic valve stenosis	14 (25%)	8 (25%)	6 (24%)	1.0
BAV	22 (39%)	15 (47%)	7 (28%)	0.18
UAV	7 (12%)	3 (9%)	4 (16%)	0.69
Coarctation	11 (19%)	7 (22%)	4 (16%)	0.74
Shone association †	5 (9%)	3 (5%)	2 (4%)	0.64
Prior surgery	32 (56%)	21 (66%)	11 (44%)	0.12
CoA repair	23 (40%)	16 (50%)	7 (28%)	0.11
VSD repair	5 (9%)	4 (13%)	1 (4%)	0.37
Other surgery	14 (25%)	9 (28%)	5 (20%)	0.55

Data are presented as medians (range) or frequency (percent). BAV, bicommissural aortic valve; BSA, body surface area; CoA, coarctation of the aorta; UAV, unicommissural aortic valve; VSD, ventricular septal defect.

* Adams-Oliver syndrome (1), Apert syndrome (1), DiGeorge syndrome (1), Down syndrome (1), hemochromatosis (1), heterotaxy (1), mosaic trisomy 20 (1), unspecified syndrome (2), Turner syndrome (1), Williams syndrome (1).

† Shone association defined as ALMR and 2 of the following: (1) parachute mitral valve, (2) sub AS, (3) coarctation.

Table 2

Echocardiographic data

	All Patients (n= 57)	ALMR Resection (n= 32)	No ALMR Resection (n= 25)	P-value
Isolated ALMR	6 (11%)	1 (3%)	5 (20%)	0.08
Initial MV gradient * (mm Hg)	6 (1 – 25)	7.5 (1 – 20.0)	5 (1 – 25.0)	0.06
Highest MV gradient * (mm Hg)	8 (1 – 16)	9 (1 – 15)	5 (1 – 16)	<0.001
Lateral MV annulus Z-Score	–0.30 (–4.00 – 2.79)	–0.5 (–4.0 – 2.5)	–0.1 (–2.6 – 2.8)	0.34
AP MV annulus Z-score	–0.09 (–3.00 – 4.60)	0.1 (–2.5 – 3.8)	–0.1 (–3.0 – 4.6)	0.87
Mitral valve morphology				
Closely spaced PMs	19 (33%)	12 (38%)	7 (28%)	0.57
Short chordae	23 (40%)	17 (53%)	6 (24%)	0.03
Parachute MV	8 (14%)	4 (13%)	4 (16%)	0.72
LA area Index (cm ² /m ²)	14.7 (4.1 to 46.1)	17.5 (6.8 to 28.6)	9.4 (4.1 to 46.1)	0.005
LV EDV Z-Score	0.60 (–2.03 – 8.25)	0.3 (–2.0 – 7.2)	0.6 (–1.1 – 8.3)	0.77
LV mass Z-Score	0.97 (–6.18 – 10.55)	0.9 (–2.1 – 4.3)	1.5 (–6.2 – 10.6)	0.45
LV ejection fraction	65 (11 – 78)	66 (50 – 77)	65 (11 – 78)	0.70

Data are presented as medians (range) or percent (%). ALMR, annuloleaflet mitral ring; AP, anterior-posterior; LA, left atrium; LVEDV, left ventricular end-diastolic volume; MV, mitral valve; PM, papillary muscle.

* Mean gradient.

Table 3

Factors associated with ALMR resection

	Odds Ratio	95% Confidence Intervals	P-Value
Mean MS Gradient ($\uparrow$ 1 mm Hg)	1.38	(1.11, 1.73)	0.005
LA Area Index ($\uparrow$ 1 mL/m ²)	1.07	(0.99, 1.1)	0.06
Short chordae (present)	3.59	(1.14, 11.3)	0.03
Earlier age at diagnostic echocardiogram ($\downarrow$ 1 year)	1.14	(1.03, 1.27)	0.02

MS, mitral stenosis; LA, left atrium.

Table 4

Surgical interventions and follow-up in 57 patients with ALMR

ALMR resection	32 (56%)
Age at ALMR resection (y)	1.5 (0 – 16.8)
Additional MV procedures *	14 (24%)
Splitting/mobilization of papillary muscles	9
Augmentation of chordal spaces	6
Leaflet augmentation	5
Commissuroplasty	4
Additional non-MV surgery *	24 (42%)
Subaortic stenosis resection	10
VSD closure	9
Coarctation repair	5
Aortic valvuloplasty	3
EFE resection	2
Other †	2
Follow-up data	
Early post-surgery MV gradient (mm Hg)	3 (1 – 9)
Latest MV gradient (mm Hg)	4.3 (1 – 17)
Age at last echocardiogram (y)	4.8 (0.4 – 76.9)
Recurrence of ALMR	4 (7%)
Repeat ALMR resection	2 (4%)
Additional non-ALMR surgery #	7 (12%)

ALMR, annuloleaflet mitral ring; MV, mitral valve; VSD, ventricular septal defect; EFE, endocardial fibroelastosis.

* Performed contemporaneously with ALMR resection

† Tetralogy of Fallot repair (1), aortic valve replacement (1)

Arch augmentation (1), coronary sinus surgery (1), mitral valvuloplasty (1), mitral valve replacement (2), and pacemaker (1)