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CASE SERIES
Congenital heart disease

Surgical outcome of isolated congenital supravalvular pulmonary stenosis: a case series

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Background Supravalvular stenosis of main pulmonary artery is a rare anomaly characterized by the presence of constriction band just above the pulmonary valve. It is mostly acquired after intervention on the pulmonary trunk or less commonly is congenital in origin associated with complex congenital cardiac malformations and very rarely can present as an isolated native congenital supravalvular pulmonary stenosis (SPS).

Case summary We present a series of four cases of isolated congenital SPS who underwent surgical correction at our tertiary care institute over 8 years. Mean age of the patients was 2.25 ± 0.96 years with all of them being males. Mean peak systolic gradient across the stenosis was 82 ± 21.48 mmHg ranging from 60 mmHg to 110 mmHg. There was no early and medium-term mortality with 100% survival at mean follow-up of 31 months (range 7–85 months).

Discussion Surgical correction of congenital SPS carries excellent early and mid-term results with almost no mortality and very low risk of re-intervention for restenosis.

Keywords Pulmonary stenosis • Supravalvular • Case series • Patch augmentation

Learning points

- Isolated congenital supravalvular pulmonary stenosis (SPS) is a very rare anomaly. It can be misdiagnosed as valvular pulmonary stenosis on transthoracic echocardiography and may require angiocardiography for confirmation.
- Surgical outcome of isolated SPS is excellent.

Introduction

Pulmonary artery stenosis can occur at any level from its origin till peripheral branches. It is more commonly seen with complex congenital cardiac malformations like tetralogy of Fallot, double outlet right ventricle (RV), transposition of great arteries, mainly at valvular, branch pulmonary artery origin, and less frequently at the peripheral intrapulmonary branches.[1] The stenosis can be discrete or more commonly at multiple levels. Isolated discrete congenital supravalvular pulmonary stenosis (SPS) is very rarely seen.[1,2] Here, we present four cases of congenital isolated SPS seen over a period of 8 years at our tertiary care institute.

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Timeline

Time	Events
Patient 1	
9 months old	Supravalvular pulmonary stenosis at the level of sinotubular junction with peak systolic gradient of 72 mmHg diagnosed on transthoracic echocardiography (TTE)
1 year old	Pericardial patch augmentation of main pulmonary artery with extension of patch into one sinus done. Immediate post-operative gradient in the pulmonary artery was 4 mmHg
Patient 2	
24 months old	Supravalvular stenosis due to a fibrous band just above the sinotubular junction with peak systolic gradient of 110 mmHg diagnosed on TTE
27 months old	Excision of the band with pericardial patch augmentation of main pulmonary artery done. Immediate post-operative gradient in the pulmonary artery was 5 mmHg
Patient 3	
1 year old	Supravalvular pulmonary stenosis at the level of sinotubular junction with peak systolic gradient of 86 mmHg diagnosed on TTE
1 year old	Attempted balloon dilatation of the stenosis but failed
3 years old	Pericardial patch augmentation of main pulmonary artery with extension of patch into two sinuses done. Immediate post-operative gradient in the pulmonary artery was 10 mmHg
Patient 4	
2 years old	Supravalvular pulmonary stenosis at the level of sinotubular junction with peak systolic gradient of 60 mmHg diagnosed on TTE
3 years old	Pericardial patch augmentation of main pulmonary artery with extension of patch into one sinus done. Immediate post-operative gradient in the pulmonary artery was 8 mmHg

Case presentation

Patient 1

Isolated SPS was diagnosed in a 1 year male child who presented with exertional dyspnoea without cyanosis. The child was haemodynamically stable with normal vitals. His cardiovascular examination revealed normal S1 and S2 with ejection systolic murmur in pulmonary area without thrill and no radiation. His respiratory and rest of the systemic examination was normal. The electrocardiogram (ECG) of the patient showed right axis deviation with features suggestive of right ventricular hypertrophy with tall R wave in right and deep S wave in left precordial leads respectively. P wave was normal. There was RV type of apex on chest X-ray (CXR) with normal pulmonary vascularity. There were no other significant findings on CXR. There was a peak systolic gradient of 72 mmHg across the stenosis on transthoracic echocardiography (TTE) without pulmonary regurgitation with hypertrophied RV. The diagnosis was confirmed on cardiac angiography (*Figure 1*). No other cardiac anomaly was detected. The stenosing ridge was present just above the sinotubular junction of the main pulmonary artery. Autologous pericardial patch augmentation of the main pulmonary artery with extension of the patch into the anterior sinus was done on cardiopulmonary bypass (CPB). The pulmonary valve was tricuspid with thickened, non-stenosing leaflets, and commissures were not fused. Post-operatively, residual peak systolic gradient was 4 mmHg.

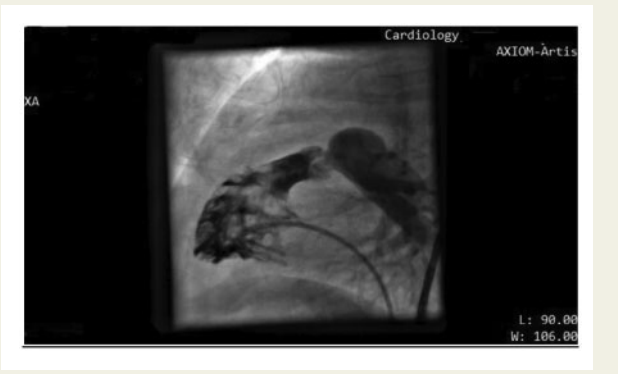

Figure 1 Supravalvular pulmonary stenosis with mild post-stenotic dilatation (lateral view).

Patient 2

Two years acyanotic, male child presented with exertional dyspnoea and palpitations. His haemodynamics were within normal limits. There was an ejection systolic murmur in pulmonary area with systolic thrill with normal S1 and S2 heart sounds. Rest of the systemic and general physical examination was normal. The ECG had features of right ventricular hypertrophy with right ventricular strain pattern on right precordial leads with right axis deviation. P wave was slightly peaked. His CXR revealed RV type of apex with normal pulmonary vascularity. His TTE revealed isolated supravalvular ridge creating a peak systolic gradient of 110 mmHg without significant post-stenotic pulmonary artery dilatation. Right ventricle was hypertrophied. There was no pulmonary regurgitation. Cardiac angiography was done for the confirmation of the diagnosis (*Figure 2*). The supravalvular fibrous band was excised and main pulmonary artery was augmented with autologous pericardial patch. Pulmonary valve was tricuspid with normal leaflets and commissures. There was a gradient of 5 mmHg in the pulmonary artery post-operatively.

Patient 3

This was a 3 years male child admitted with exertional dyspnoea without cyanosis. His vitals were normal for his age. The child had

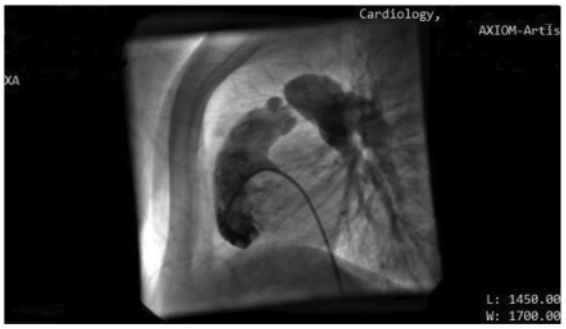

Figure 2 Severe supravalvular pulmonary stenosis due to a fibrous band (lateral view).

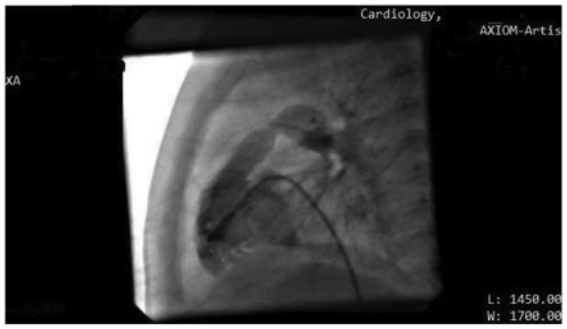

Figure 3 Supravalvular pulmonary stenosis post failed balloon dilatation (lateral view).

normal heart sounds with normal respiratory variations with only an ejection systolic murmur in pulmonary area without thrill. His rest of the general physical and systemic examination had no significant findings. There were features suggestive of right ventricular hypertrophy with right axis deviation on ECG. Right atrial P wave appeared prominent on Lead II. The pulmonary vascularity appeared normal with RV type of cardiac apex on his CXR. His TTE revealed severe SPS with peak systolic gradient of 86 mmHg with no pulmonary regurgitation with hypertrophied RV. There was a small 4 mm patent foramen ovale (PFO) with left to right shunting. No other cardiac lesion was identified. The child had undergone failed attempt at balloon dilatation of the supravalvular stenosis at the age of 1 year (*Figure 3*). The child was operated electively and was found to have a constricted segment causing isolated SPS just above the pulmonary valve. The main pulmonary artery was augmented using autologous pericardial patch extending into the two sinuses. The pulmonary valve was tricuspid with thickened leaflets. Small PFO was closed by direct suturing. A residual gradient of 10 mmHg was present across the pulmonary valve post-operatively.

Patient 4

In a 3 years acyanotic, male child, presenting with recurrent chest infections and exertional dyspnoea, severe pulmonary stenosis (PS) was diagnosed on TTE with peak systolic gradient of 60 mmHg and a small 3 mm PFO with left to right shunting without any other cardiac lesion. Pulmonary regurgitation was absent. The heart rate and blood pressure of the child was within normal limits for his age. The S1 and S2 heart sounds were normal in intensity with normal respiratory variations. There was a non-radiating, ejection systolic murmur in left second intercostal space without thrill. The rest of the systemic examination had no significant findings. Right ventricular hypertrophy features were evident on ECG along with right axis deviation. P wave was normal. The enlarged RV occupied the apex on CXR with normal pulmonary vascularity. The diagnosis was confirmed on cardiac angiography (*Figure 4*). The child was found to have isolated SPS at the sinotubular junction. Pulmonary valve was tricuspid and leaflets minimally thickened with mild tethering. Right ventricle was hypertrophied. Autologous pericardial patch augmentation of the main

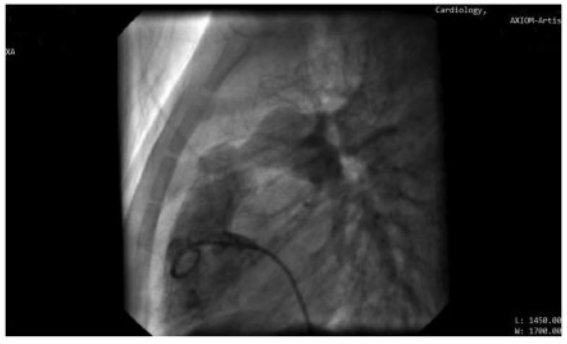

Figure 4 Supravalvular pulmonary stenosis with mild pulmonary valve tethering (lateral view).

pulmonary artery and one anterior sinus was done. Small PFO was closed by direct suturing. Post-operative gradient across the pulmonary valve was 8 mmHg.

Table 1 illustrates the clinical characteristics of the patients. Mean age of the patients was 2.25 ± 0.96 years with all of them being males. Mean body surface area was 0.53 ± 0.04 m^2. Mean peak systolic gradient across the stenosis was 82 ± 21.48 mmHg ranging from 60 mmHg to 110 mmHg. All the patients were done on CPB on arrested heart except Patient 4 which was done on beating heart. Mean aortic cross-clamp time was 59.67 ± 35.92 min (range 36–101 min) and CPB time was 81 ± 38.03 min (range 48–135 min). Patients were mechanically ventilated for an average of 8.48 ± 4.91 h (range 3.75–15.33 h). Median intensive care unit stay was 3 days (range 1–6 days) and median hospital stay was 6.5 days (range 4–11 days). There was no early and medium-term mortality with 100% survival at mean follow-up of 31 months (range 7–85 months). All the patients showed marked symptomatic improvement. All the patients were in New York Heart Association (NYHA) I class till last follow-up with normal right ventricular function. Patients will be followed-up annually and have TTE every 2–3 years or earlier if there is any change in symptoms.

Table I Clinical characteristics of the patients

Patient number	Age (years)	Sex	BSA (m^2)	AXC time (min)	CPB time (min)	ICU stay (days)	Hospital stay (days)	Peak GR. (pre-op)	Peak GR. (post-op)	Peak GR. (follow-up)	Associated anomalies	Duration of MV (h)	F/U (months)
1.	1	M	0.48	42	63	2	4	72	4	5	None	8	85
2.	2	M	0.52	101	135	4	6	110	5	13	None	15.33	20
3.	3	M	0.54	36	78	6	11	86	10	28	4MM PFO P/BPV in 2015	6.83	11
4.	3	M	0.58	Beating heart	48	1	7	60	8	9	3MM PFO	3.75	7

AXC, aortic cross-clamp; BSA, body surface area; CPB, cardiopulmonary bypass; F/U, follow-up; ICU, intensive care unit; M, male; MV, mechanical ventilation; peak GR., peak systolic gradient across the stenosis; pre-op, pre-operative; post-op, post-operative.

Discussion

Supravalvular pulmonary stenosis is a rare anomaly characterized by the stenosis of main, central branch, or peripheral branch pulmonary arteries either in combination or as an isolated lesion and is more commonly seen with congenital rubella syndrome.[3] Multiple level stenoses is more common than an isolated obstruction. Isolated SPS is more commonly acquired, and occurs after surgical intervention involving the main pulmonary artery.[4–6] Isolated congenital SPS is the rarest form of pulmonary stenosis.[1,2] The rarity of the isolated congenital SPS can be gauged from the study done by Yuan.[7] who reviewed literature for the SPS, congenital, and acquired, from 2000 to 2016 and found only 4 out of 333 (1.2%) cases of isolated congenital SPS. We also encountered only 4 (0.08%) cases out of 4231 surgeries done for congenital cardiac disease at our tertiary care institute over 8 years. The presence of a hypertrophied stenosing ring of tissue at the sinotubular junction of the main pulmonary artery is its characteristic feature. The pulmonary valves are most of the times thickened depending upon the degree of stenosis because of the persistent trauma by the blood jet. It is usually misdiagnosed as pulmonary valvular stenosis on TTE and usually confirmed on cardiac catheterization and pulmonary angiography study. There are few individual case reports[8–11] but only two case series so far to the best of our knowledge.[12,13] Bacha *et al.*[12] reported surgical autologous tissue repair of SPS in eight cases. However, only four of them were isolated SPS and rest of the four cases were associated with complex congenital cardiac malformations. They also reported zero early and medium-term mortality without any re-intervention over a follow-up of 22 months. Dogan *et al.*[13] emphasized the use of oval pericardial or Dacron patch repair of isolated SPS in their experience of four cases with zero mortality. The presenting age of the patients is variable depending upon the severity of the stenosis. We encountered the disease in toddlers but there have been case reports of presentation in neonates,[8] children,[11] and even in a septuagenarian.[9]

The severity of SPS is classified on the basis of peak systolic gradient on Doppler ultrasonography or pressure gradient across the stenosis during cardiac catheterization study, similar to valvular PS, as mild (<30 mmHg), moderate (30–50 mmHg), and severe (>50 mmHg) as per American Heart Association guidelines.[14] Surgery is recommended for pulmonary stenosis when the patient becomes symptomatic or if the stenosis is severe. There is no role for medical therapy. Percutaneous intervention may be tried, but surgery is the mainstay of SPS management in contrast to pulmonary valvular stenosis where balloon valvotomy is the first line of therapy.[14] There are multiple management strategies for dealing with SPS. Balloon dilatation/stent implantation is most often tried initially[15,16] but fails most of the times because of the elastic recoil of the supravalvular ridge.[7] Stenting is avoided due to the proximity of the valve and risk of stent thrombosis or stent migration. Surgical techniques include simple resection of the stenotic tissue followed by anastomosis to incising the stenotic area and then patching it with a single or multiple patches or using an autologous tissue for the augmentation of the stenotic area.[1,6,12] Considering the rarity of the isolated congenital SPS, long-term follow-up data are scarce, and therefore, definite conclusions on the re-intervention rates cannot be drawn. However, none of the series in the literature so far have reported any re-intervention.[12,13]

Conclusion

Whatever be the technique, main aim is to relieve the stenosis and decrease the risk of re-stenosis. The diagnosis of pulmonary stenosis whether valvular or supravalvular is important from management aspect as balloon valvotomy is the primary treatment modality for the former, while surgery is the mainstay for the latter. The surgical results, both early and medium-term, of isolated SPS are extremely good and without any mortality.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patients in line with COPE guidance.

Conflict of interest: none declared.

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