Prognostic à long terme des enfants opérés pour syndrome de Shone

Study of cardiac function in 2d strain in healthy children

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Résumé

Introduction : Le syndrome de Shone (SSh) est compsé d'obstructions étagées du cœur gauche. Malgré la progression de la chirurgie, les patients peuvent présenter des récidives d'obstructions ou de complications de prothèses nécessitant une réintervention.

Observations : Nous présentons une série de cas de patients traités chirurgicalement pour un SSh complet et présentant un mauvais pronostic à long terme. Nous avons cherché à déterminer les résultats à long terme, la mortalité et la nécessité de réinterventions après la chirurgie du SSh complet.

Conclusion : le SSh est associé à un mauvais pronostic à long terme.

Mots-clés

Syndrome de Shone, chirurgie cardiaque, pronostic

Summary

Introduction : Shone's syndrome (ShS) is composed of various left heart tract obstructions. Despite the improvements in surgical strategies, patients may present recurrent obstructions or prostheses complications at the long-term follow-up, which requires re-intervention.

Observation: we, herein, present case series of patients treated surgically for complete ShS and presenting poor long-term prognosis to determine the long-term outcomes, mortality, and the need of re-interventions after complete ShS surgery.

Conclusion: ShS is associated with poor long-term prognosis and the need of re-intervention.

Keywords

Shone syndrome, cardiac surgery, prognosis

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INTRODUCTION

Shone syndrome (ShS), first described by Dr. John D. Shone in 1963, is defined by multilevel congenital left heart outflow tract obstruction [1]. It accounts for less than 1% of all congenital heart diseases [1]. Complete ShS includes various left-sided cardiac defects, namely mitral supravalvular membrane, parachute mitral valve, subaortic membrane, bicuspid aortic valve, and coarctation of the aorta (CoA) [2]. Transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), and cardiac computed tomography scan (CTS) play a crucial role in the diagnosis and in guiding the therapeutic strategies. Surgical treatment remains challenging [3]. Some reports have focused on the surgical outcomes demonstrating the need of repeated re-intervention . We, herein, present case series of five patients suffering from complete ShS with different initial clinical presentations, managements, and long- term evolution. The aim of this study was to highlight the outcomes, mortality, and the need of re-interventions after complete ShS surgery.

OBSERVATIONS

All the patients admitted for complete ShS were retrospectively enrolled between 2000 and 2023. Long term follow-up was reported for all the patients. TTE, TEE, and cardiac CTS were performed to identify the need of re-intervention on valvular deterioration or recoarctation. An oral consent was obtained from the patients or the families of the dead patients.

Case 1

A 1-year-old female child presented with recurrent respiratory infection. Preoperative TEE revealed hypertrophic left ventricle (LV) with normal ejection fraction (EF = 60%–65%), enlarged left atrium (LA), obstructive supra mitral ring (mean gradient = 24 mmHg) with parachute mitral valve (figure 1), bicuspid aortic valve with mild aortic regurgitation, obstructive subaortic membrane (peak gradient =50mmHg), and CoA. A single-stage repair was carried out through median sternotomy including resection of CoA with end-to-end anastomosis of descending thoracic aorta followed by resection of supra mitral ring, splitting of papillary muscle and subaortic membrane resection. Long term evolution was marked by the recurrence of severe mitral stenosis requiring surgical mitral valve replacement by mechanical prosthesis. Three weeks after the surgery, the patient died of a severe hemorrhagic stroke.

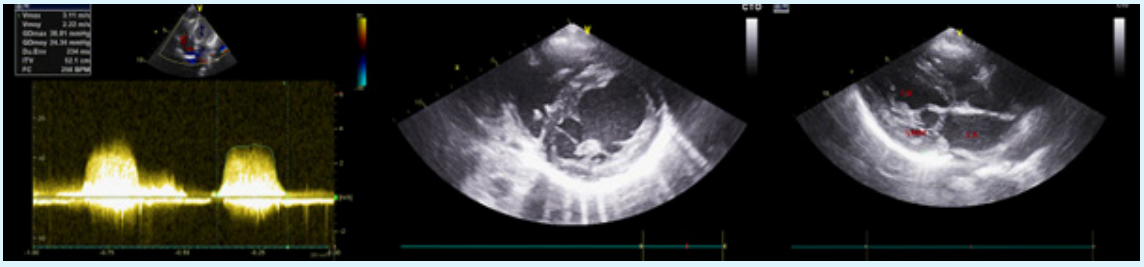

Figure 1. Multilevel mitral obstructions on Trans-thoracic echocardiography: supra mitral ring with mean gradient = 24 mmHg and parachute mitral valve. LA : left atrium, LV : left ventricle, SMM : supra mitral membrane.

Case 2

A newborn male patient presented with cardiogenic shock and severe dyspnea. Preoperative TTE revealed nonobstructive mitral ring (mean gradient 5 mmHg), single papillary muscle, nonobstructive subaortic membrane, severe CoA (peak gradient = 65mmHg), and altered LV function. The patient underwent surgical resection of CoA and end-to-end anastomosis of descending thoracic aorta through lateral thoracotomy approach. A four-year follow-up was marked by the aggravation of mitral valve stenosis (mean gradient= 16mmHg), leading to surgical mitral valve replacement. At the age of 18 years, acute mitral prosthesis obstruction occurred due to infective endocarditis. Urgent surgery was performed; however, the patient died immediately after the surgery.

Case 3

A 7-year-old female patient presented with dyspnea on exertion. The clinical examination revealed 150/100 with reduced pulse in the lower limbs. TTE revealed moderate multilevel left-sided obstruction associated with severe CoA. Resection of CoA was performed. Evolution was marked by severe hypertension and recoarctation, confirmed by aorta CT, requiring aorta stenting at the age of 20 years. At the age of 30 years, the evolution was marked by aorta stent stenosis (figure 2) inducing severe hypertension and necessitating stent dialation by balloon.

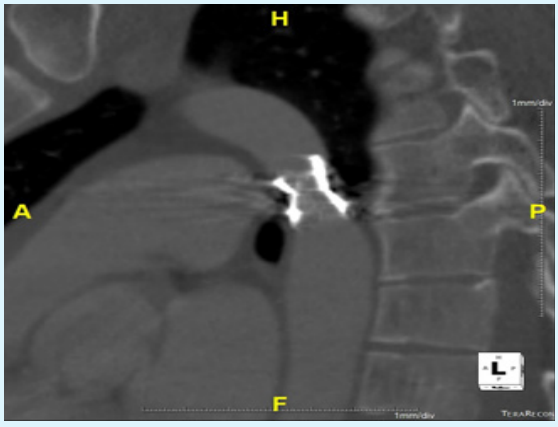

Figure 2 : Saggital view on the aorta CT scan: stent stenosis and post- stenosis dilatation of the descending aorta.

Case 4

A 12-year-old female child presented with NYHA III class III of dyspnea. Preoperative TTE revealed severe mitral stenosis (mean gradient= 16 mmHg) due to supra mitral ring and parachute mitral valve associated with bicuspid aortic valve, mild aortic regurgitation, severe subaortic membrane obstruction (mean gradient = 52 mmHg), and mild LV hypertrophy with normal function. The patient underwent mitro-aortic valves replacement using mechanical prostheses, with a good postoperative evolution. At the 6-year follow-up, the patient presented severe mitral prosthesis stenosis, LA dilatation, no aortic prosthesis stenosis, and severe tricuspid regurgitation with annular dilatation. The patient underwent re-intervention consisting of mitral prosthesis replacement and tricuspid valve annuloplasty, with a good postoperative evolution.

Case 5

A two-month-old female patient presented with severe dyspnea and difficulty in breastfeeding. Preoperative TTE revealed nonobstructive mitral ring, parachute mitral valve, severe obstructive subaortic membrane, bicuspid aortic valve, and moderate CoA (peak gradient = 30 mmHg) with hyperkinetic LV. The patient underwent surgical resection of the subaortic membrane, with a good evolution. The six-year follow-up was marked by the aggravation of mitral valve stenosis associated with severe mitral regurgitation, LV dilatation, and aorta coarctation with no aortic valve obstruction. The patient underwent mitral valve replacement using mechanical prosthesis and resection of coarctation, with an initial good evolution under oral anticoagulation. At the age of sixteen, she had anticoagulation overdose complicated by cerebral bleeding and brain herniation.

DISCUSSION

ShS is a rare congenital condition of obstructive lesions of the left heart outflow tract [1]. It is manifested with a large spectrum of symptoms depending on the severity of the defects. Diagnosis is usually made during childhood. The age of the symptoms' onset depends on the form, whether complete or incomplete, and the severity of LV inflow and outflow obstructions [2]. A combination of the imaging results (echocardiography and CT scan) is essential for an accurate diagnosis and an appropriate treatment choice [4]. The therapeutic strategies are multiple and the outcomes are not well-reported. Few surgical series have reported the long-term postoperative survival rates. Brown et al. reported a five-year survival rate of 93% [5]. The poorest prognosis has often been related to mitral obstruction severity and pulmonary hypertension. Studies have reported that mitral valve repair has a better long-term prognosis compared to valve replacement [6]. The valve prosthesis does not allow mitral annular growth and it needs lifelong anticoagulation. Many patients experience recurrent LV inflow and outflow dysfunction, leading to an increased rate of re-intervention and poor long-term prognosis.

CONCLUSION

The surgical treatment of complete ShS should be individualized according to the lesions' severity. Severe mitral stenosis and coarctation of the aorta are associated with a high likelihood for re-interventions.

What is already known on this topic?

- Complex Shone syndrome is characterized by clinical polymorphism

- The complications are varied and could be lethal

- Repeated cardiac surgery could be indicated

What this study adds.

- Worse prognosis of operated Shone syndrome in adults

- The benefits of cardiac catheterization in Shone syndrome

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