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Balancing Risks and Benefits: Perioperative Management of Severe Pulmonary Stenosis in Noncardiac Surgery - A Case Study

Equilibrando riesgos y beneficios: Manejo perioperatorio de la estenosis pulmonar grave en cirugía no cardíaca – Un estudio de caso

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SUMMARY

Background: *Pulmonary stenosis accounts for 7 %-10 % of all congenital heart disease cases. Given their extended lifespans, these patients often require increased diagnostic, interventional, and surgical interventions for noncardiac conditions. This case report focuses on the perioperative evaluation of a patient diagnosed with severe pulmonary stenosis who was scheduled for non-cardiac surgery.* **Case Presentation:** *A 19-year-old girl was referred to the Cardiology Department for a preoperative assessment before undergoing Salpingo-Oophorectomy for an Ovarian Cystic Neoplasm. Since the age of 6, she has experienced limited exercise tolerance and exertional dyspnea. Echocardiography revealed severe pulmonary valve stenosis. Following cardiac catheterization and balloon pulmonary valvuloplasty (BPV), the patient underwent a left-sided Salpingo-Oophorectomy without any complications. She was discharged home three days after the procedure.* **Conclusions:** *Patients with congenital heart disease require thorough evaluation by a multidisciplinary team of experts before surgery, with meticulous planning. This case underscores the significance of perioperative assessment and care in patients with severe pulmonary stenosis undergoing noncardiac surgery.*

Keywords: *Pulmonary stenosis, perioperative, balloon pulmonary valvuloplasty, noncardiac surgery.*

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RESUMEN

Introducción: *La estenosis pulmonar representa del 7 % al 10 % de todos los casos de enfermedad cardíaca congénita. Dada su esperanza de vida prolongada, estos pacientes a menudo requieren intervenciones diagnósticas, intervencionistas y quirúrgicas aumentadas para condiciones no cardíacas. Este informe de caso se centra en la evaluación perioperatoria de una paciente diagnosticada con estenosis pulmonar grave que estaba programada para cirugía no cardíaca.* **Presentación del Caso:** *Se remitió a una joven de 19 años al Departamento de Cardiología para una evaluación preoperatoria antes de someterse a una Salpingo-Ooforectomía por un Neoplasma Quístico Ovárico. Desde los 6 años de edad, había experimentado una tolerancia al ejercicio limitada y disnea de esfuerzo. La ecocardiografía reveló una estenosis grave de la válvula pulmonar Tras una cateterización cardíaca y valvuloplastia pulmonar con balón (VPB), la paciente se sometió a una Salpingo-Ooforectomía del lado izquierdo sin complicaciones. Fue dada de alta a su hogar tres días después del procedimiento.* **Conclusión:** *Los pacientes con enfermedad cardíaca congénita requieren una evaluación exhaustiva por parte de un equipo multidisciplinario de expertos antes de la cirugía, con una planificación meticulosa. Este caso subraya la importancia de la evaluación y el cuidado perioperatorio en pacientes con estenosis pulmonar grave que se someten a cirugía no cardíaca.*

Palabras clave: *Estenosis pulmonar, perioperatorio, valvuloplastia pulmonar con balón, cirugía no cardíaca.*

BACKGROUND

Congenital heart disease (CHD) comprises structural and functional disease of the heart, with an incidence of nearly 1 % of births in the United States. The leading cause of death between 1999 and 2006 was CHD in 27 960 people, with 48 % of fatalities being infants due to complex surgeries and prolonged intensive care unit (ICU) stays (2). As the population of adults with congenital heart disease grows, they will likely present for non-cardiac procedures in increasing numbers (1). Risk stratification and an interdisciplinary approach are paramount in achieving safe outcomes. Perioperative risk in adults with CHD is more complex to evaluate due to the variable expression of CHD and the superimposed cardiac disease.

In the realm of noncardiac surgery planning, the presence of congenital valvular heart disease poses a potential challenge for cardiologists. Pulmonary stenosis, accounting for 7 %-10 % of total congenital heart disease, demonstrates a slight female predominance (3). This case report underscores the importance of perioperative evaluation in a patient diagnosed with severe pulmonary stenosis scheduled for non-cardiac surgery.

Case presentation

A 19-year-old girl was referred to the Cardiology Department for perioperative evaluation in preparation for a Salpingo-Oophorectomy for Ovarian Cystic Neoplasm. Congenital heart disease was suspected in the patient. Since the age of 6 years, she has experienced limited exercise tolerance and exertional dyspnea, with no history of cyanosis.

Clinical examination revealed the patient lying flat in bed, with a heart rate of 90 beats per minute, a baseline blood pressure of 115/70 mmHg, and a respiratory rate of 20 breaths per minute. Auscultation detected a systolic crescendo-decrescendo ejection murmur at the left upper sternal border, along with soft and delayed P2 and normal S1 and widely split S2 heart sounds. Electrocardiography demonstrated sinus rhythm, right axis deviation, and right ventricular hypertrophy (Figure 1). Laboratory findings revealed leukocytosis (13 600), normocytic normochromic anemia (hemoglobin 10.2 g/dL), and hypoalbuminemia (2.7 g/dL). Meanwhile, chest X-ray findings revealed levocardia, pulmonary plethora, enlarged main pulmonary artery, and right ventricular hypertrophy (Figure 2).

Her echocardiogram findings showed a stenotic, dooming annulus (15 mm) with severe pulmonary stenosis with a pulmonary pressure gradient of 100 mmHg. A stretched patent foramen ovale, severe tricuspid regurgitation, and right ventricle dilatation also were observed (Figure 3)..

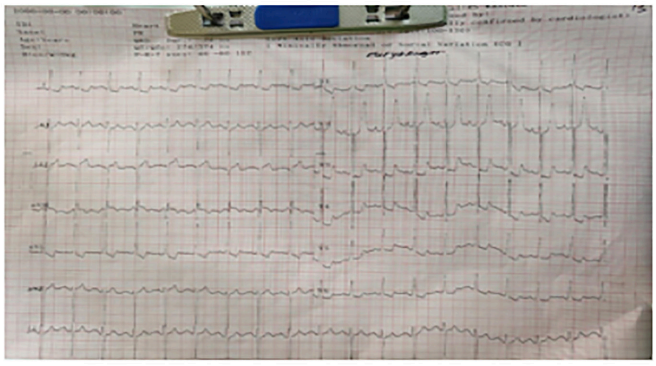

Figure 1 . Electrocardiography showed sinus rhythm, right axis deviation, and right ventricular hypertrophy.

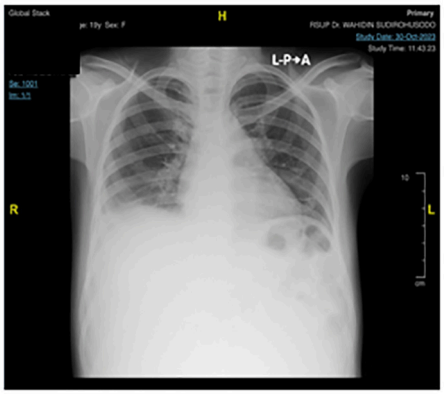

Figure 2 . Chest X-ray showed levocardia, pulmonary plethora, enlargement of the main pulmonary artery, and right ventricular hypertrophy.

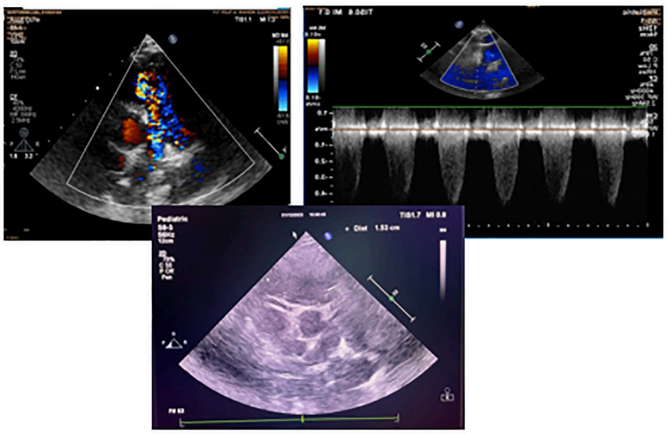

Figure 3. Echocardiography revealed a severe pulmonary stenosis with a pressure gradient of 100 mmHg and severe tricuspid regurgitation.

Subsequently, the patient underwent cardiac catheterization and balloon pulmonary valvuloplasty. The right heart catheterization revealed severe pulmonary valvar stenosis, with a right ventricle to pulmonary artery (RV-PA) gradient of 125 mmHg and a pulmonary artery annulus measuring between 15.8 mm and 16.7 mm. Following this, the decision was made to perform balloon pulmonary valvuloplasty using a Balloon Nucleus with a diameter of 20 mm x 40 mm. Evaluation after the procedure showed a reduction in the RV-PA gradient to 100 mmHg (Figure 4).

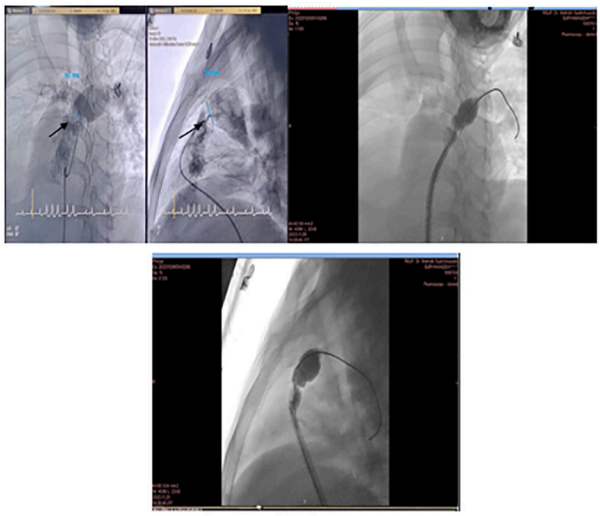

Figure 4. Right Heart Catheterization and Ballon Pulmonary Valvuloplasty. Annulus Pulmonary Artery *(black arrow)*.

One day after balloon pulmonary valvuloplasty (BPV), an echocardiography evaluation revealed a pulmonary stenosis gradient of 60 mmHg. Seven days after BPV, the patient underwent a left salpingo-oophorectomy procedure, which lasted for 2 hours without complications. The amount of bleeding was recorded as 750 mL. Following the procedure, the patient was transferred to the ward for observation. Subsequently, the patient was discharged home three days after surgery.

DISCUSSION

Advancements in pediatric cardiology, cardiac surgery, anesthesia, and critical care have significantly enhanced the survival rates of patients with congenital heart disease (CHD). With the aging population, there is a growing likelihood of an increasing number of individuals requiring non-cardiac surgeries (1). Pulmonary stenosis is characterized by the stiffening of the pulmonic valve, leading to obstructed blood flow. Isolated pulmonary stenosis can be classified into valvular, subvalvular, and supravalvular obstruction. Many patients with pulmonary stenosis remain asymptomatic, potentially resulting in delayed diagnosis until an event precipitates further investigation (3).

Echocardiography and Doppler flow assessments aid in identifying the location of the obstruction and assessing the severity of stenosis. Per the 2020 ESC guidelines on adult CHD management, pulmonary stenosis (PS) can be categorized as follows: (1) Severe stenosis: Peak gradient $\geq$ 64 mmHg; (2) Moderate stenosis: Peak gradient ranging from 36 to 64 mmHg; and Mild stenosis: Peak gradient $\leq$ 36 mmHg (4).

Cardiac catheterization and echocardiography data play crucial roles in diagnosing pathophysiology and formulating surgical and anesthetic strategies. The information obtained from cardiac catheterization includes the identification of anatomical defects; saturation data for Qp: Qs ratio, shunting, and calculations for V/Q mismatch; comparison of pressures in the right and left cardiac chambers, systemic and pulmonary arteries, as well as the measurement of valve and shunt pressure gradients; and angiographic data assessing ventricular wall motion and blood flow in great vessels and cardiac chambers (4).

The 2008 ACC/AHA guidelines for the care of individuals with congenital heart disease categorize adult CHD into three risk categories: low, moderate, or high (Table 1). Low-risk lesions encompass congenital heart abnormalities that are deemed uncomplicated. Patients classified as moderate or high-risk should receive care at specialized centers, irrespective of the nature of the required surgery (1).

Mossad devised a point-based system to stratify the risk of congenital heart disease patients before surgery (Table 2). A total score of 0–6 points suggests a low perianesthesia risk, allowing for outpatient interventions. Scores ranging from 6 to 13 points indicate significant perianesthesia risk for same-day procedures, although consideration for transfer to a CHD center may be warranted. A cumulative score of 14-20 points signifies high perianesthesia risk, necessitating procedures performed in specialized centers with potential critical care admission. Preoperative cardiac catheterization benefits most high-risk patients (14-20 points). Our patient fell into the moderate risk based on lesion complexity and risk (1). Based on the point system for stratifying the risk of patients undergoing surgery, the patient received point 7.

Table 1. Classification of lesion complexity and risk in Congenital Heart Disease

Low Risk	Moderate Risk	High Risk
Small atrial septal defect	Anomalous pulmonary venous drainage, partial or total	Conduits, valved or non-valved
Isolated small ventricular septal defect (no associated lesions)	Atrioventricular septal defects (partial or complete)	Cyanotic congenital heart (all forms)
Small patent ductus arteriosus	Coarctation (CoA) of the aorta	Double-outlet ventricle
Repaired patent ductus arteriosus, secundum or sinus venosus atrial septal defect (without residua), or ventricular septal defect (without residua)	Ebstein's anomaly	Eisenmenger syndrome
	Pulmonary valve regurgitation (moderate to severe)	Fontan procedure
	Pulmonary valve stenosis (moderate to severe)	Mitral atresia
	Valvular aortic stenosis (AS) or supra AS, except hypertrophic obstructive cardiomyopathy (HOCM)	Single Ventricle
	Tetralogy of Fallot	Pulmonary atresia (all forms)
	Ventricular septal defect with other cardiac lesions	Transposition of the great arteries
		Tricuspid atresia
		Truncus arteriousus

Balloon pulmonary valvuloplasty (BPV) is indicated when the peak Doppler gradient exceeds 60 mmHg in asymptomatic individuals or 50 mmHg in symptomatic patients, serving as the primary treatment for "doming" valvular pulmonary stenosis (PS). As per the 2018 American Heart Association/American College of Cardiology (AHA/ACC) guideline, BPV is recommended for adults with moderate or severe valvar PS and otherwise unexplained symptoms of heart failure, cyanosis resulting from interatrial right-to-left communication, and exercise intolerance (class I, level of evidence B) (4,5).

Table 2 . Scoring system for assessing Adult Congenital Heart Disease patients (2).

	0	1	2
CHD diagnosis	Simple (ASD)	Moderate (ASD + PS)	Severe (TOF)
CHD interventions	Corrected, no residual	Corrected with residual	Paliated
Ventricular obstruction	None	Yes, graduent <49 mmHg	Yes, gradient >50 mmHg
Ventricular, number, and position	2 LV is systemic ventricle	1LV is systemic ventricle	1RV is systemic ventricle
Dysfunction systemic ventricle	Mild	Moderate	Severe
PVR	Normal <2 wood units	2-4 woods units	>4 woods units
SAO_2	>90 %	75 %-90 %	<75 %
Hct	30 %-45 %	25 %-30 %. or 45 %-65 %	>65 %
Arrhythmias	Seldom	Atrial ñevel	Ventricular level
Nº of cardiac medications	1	2	3

Before undergoing balloon pulmonary valvuloplasty (BPV), the patient in our case presented with extensive pulmonary stenosis, with a peak gradient measuring 100 mmHg. Following valvuloplasty, before surgery, her maximal gradient reduced to 60 mm Hg, indicating a classification of moderate stenosis. While the pressure gradient typically decreases rapidly after valvuloplasty, the precise duration required for optimal functionality and determining a safe threshold for pulmonary stenosis gradients during noncardiac surgical procedures remains unclear.

The objectives of hemodynamic management encompass maintaining optimal levels of right ventricular preload, left ventricular afterload, and right ventricular contractility. Ideally, the heart rate should remain below average. Pulmonary stenosis elevates pressure within the heart's ventricles, increasing the workload on the right ventricle. Anesthetic management primarily aims to reduce both pulmonary vascular resistance and systemic vascular resistance. It is advisable to prevent hypothermia, hypercarbia, acidosis, hypoxia, and high ventilator pressure (6).

CONCLUSIONS

The case underscores the critical necessity for comprehensive evaluation and meticulous perioperative management in patients with congenital heart disease, particularly severe pulmonary stenosis, before noncardiac surgery. Collaborative assessment by multidisciplinary teams comprising experts from various fields is imperative to ensure optimal outcomes.

DECLARATIONS

Ethics approval and consent to participate. Not applicable.

Consent for publication

The authors certify that the patient and family have obtained all appropriate patient consent forms. The patient's parents understand that names and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

Availability of data and material

Not applicable

Competing interests

The authors declare no competing interests.

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Authors' contributions

JK, AAU, and YP contributed to the initial concept and ideas. JK prepared the initial manuscript. AQ revised and prepared the submitted manuscript. AAU, YP, IM, and MZ reviewed and advised for critical revisions. All contributing authors approved the final draft.

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