

Assessment of operability in d-transposition of great arteries with ventricular septal defect: A practical method

Pankaj Bajpai, Sejal Shah, Amit Misri, Shekhar Rao, PV Suresh, Sunita Maheshwari

Department of Pediatric Cardiology, Narayana Hrudayalaya, Bangalore, Karnataka, India

ABSTRACT

- Introduction :** Pulmonary vascular disease is a risk factor in the surgical management of patients with d-transposition of great arteries (d-TGA) and a ventricular septal defect (VSD). In older infants or children with this physiology, the question of operability often arises. Cardiac catheterization in this condition can be fallacious. It is well known that oxygen reduces pulmonary arterial pressure and pulmonary vascular resistance especially where irreversible pulmonary vascular obstructive disease has still not set in. We tried to implement this effect of oxygen in correlation with echocardiography in patients with TGA-VSD physiology where operability was in question.
- Methods :** Patients with d-TGA and a large post tricuspid shunt in whom operability was considered doubtful were selected for the study. We administered humidified oxygen at the rate of 10 litres/minute by mask for 48 hours in the ward or intensive care unit. After administration of oxygen we reassessed the child echocardiographically looking for signs of lowering of pulmonary vascular resistance which included increased pulmonary venous blood flow to the left atrium (LA) and right to left shunting across the VSD.
- Observation :** We studied 1 patient with d-TGA and aortopulmonary window (APW), 4 patients with TGA / large VSD and 1 patient with Taussig-Bing anomaly. The age of the studied children ranged from 4 months to 3 years with a mean age of 1.1 years. After administering oxygen as described, echocardiogram showed an increase in pulmonary venous blood flow to the LA and right to left shunting across the VSD in 5 patients and increased flow reversal in aorta in presence of the APW.
- Conclusion :** Patients with TGA/VSD physiology with doubtful operability can be subjected to this method of determining operability using echocardiography after administering oxygen. Although not 100% accurate in predicting long term postoperative pulmonary hypertension, this is a simple, noninvasive method that can aid in decision making in such a situation.
- Keywords :** Operability, oxygen administration, reversibility testing, TGA

INTRODUCTION

Accelerated pulmonary vascular disease is prevalent in complete transposition of great arteries (d-TGA),

especially in the presence of a nonrestrictive ventricular septal defect (VSD)^[1-5] or large patent ductus arteriosus (PDA).^[6] Grade 3 or 4 Heath-Edwards histological changes are found in about 20% of infants before 2 months of life and in approximately 80% of patients after 1 year of age.^[5,7] Pulmonary vascular disease is a risk factor for the surgical management of patients with d-TGA and a VSD or any other large post-tricuspid shunt. Surgical repair, therefore, is performed in early infancy, before irreversible changes occur in the pulmonary vasculature. In older infants or children who present with this condition, the question of operability often arises. Controversy exists as to the usefulness of cardiac

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Address for correspondence: Dr. Sunita Maheshwari, Department of Pediatric Cardiology, Narayana Hrudayalaya, No. 258/A, Bommasandra Industrial Area, Anekal Taluk, Bangalore – 560 099, Karnataka, India. E-mail: sunita.maheshwari@telradsol.com

catheterization in this particular situation. Cardiac catheterization to determine operability in patients with d-TGA-VSD physiology and severe pulmonary artery hypertension has certain fallacies such as pulmonary artery saturation being higher than that of the aorta due to parallel circulation, and difficulty in entering the pulmonary artery for pressure measurements. Additionally, cardiac catheterization, being an invasive procedure, has certain risks and complications in these cyanotic, ill, and pulmonary hypertensive patients. There are no clear guidelines, therefore, as to who should be catheterized. In order to assess operability without a cardiac catheterization in this situation, we developed a simple screening test, using inhaled oxygen and postoxyggen echocardiography findings, to help make this decision.

METHODS

We selected patients with d-TGA with a large post-tricuspid shunt, e.g., VSD, PDA, aortopulmonary window (APW), who had presented to the hospital late and were clinically judged to be worrisome for operability based on the following findings: (a) oxygen saturation below 80–85%; (b) a quiet precordium and no clinical signs of increased flow; (c) chest X-ray without plethora or cardiomegaly; (d) an echocardiogram showing a predominantly left to right shunt across the VSD suggesting that pulmonary vascular resistance (PVR) was higher than systemic vascular resistance (SVR) and

absence of left heart dilation in addition to the reduced pulmonary venous return.

In this subgroup of patients, after taking parental consent and informing them of our clinical dilemma, we administered heated and humidified oxygen at the rate of 10 l/min by appropriate-sized masks for 48 h (not strictly continuous) in the ward or Intensive Care Unit (ICU). After the administration of oxygen, we reassessed the child echocardiographically paying special attention to signs of an increased pulmonary blood flow suggestive of a fallen PVR by:

1. color Doppler with (a) increased pulmonary venous blood flow to the left atrium (b) right to left shunting across the VSD, left to right shunting across the PDA or increased flow reversal in aorta in presence of AP window
2. 2-D echo showing presence of dilatation of left atrium (LA) and left ventricle (LV).

RESULTS

We present herewith a summary of the result of these cases fulfilling the eligibility criteria in a tabular form in whom we applied our clinical method of determining operability [Table 1].

In this clinical series, a total of six patients were administered oxygen to assess operability, with an age range of 4 months to 3 years (mean age: 1.1 years). Out of six patients studied, five patients appeared operable

Table 1: The effect of oxygen on echo preoperatively and surgical outcome in patients with TGA and a large post-tricuspid shunt

S. no.	Age	Preoxygen echo findings	Echo after O ₂ inhalation	Surgery performed	Echo before discharge
1	4 months	d-TGA, intact IAS/IVS, large APW, no reversal of flow in the arch, severe PAH	Flow reversal in the descending aorta	ASO + APW repair	Trivial TR with a gradient of 25 mmHg. No residual AP window. Normal left ventricular function
2	13 months	Taussig–Bing anomaly	Increased pulmonary venous return	ASO + multiple VSD's closure	Mild TR with a gradient of 50 mmHg. 2–3 tiny mid muscular VSD's shunting bi-directional. Good ventricular function
3	3 years 2 months	d-TGA with I malposed great arteries, a large subpulmonic VSD predominantly L-R shunt, additional mid muscular VSD, severe PAH	VSD demonstrated a R-L shunt. Increased pulmonary venous return	Senning's operation + multiple VSD's closure	Mild TR with a gradient of 30 mmHg; unobstructed baffles. Good ventricular function
4	6.5 months	d-TGA, a large subpulmonic VSD predominantly L-R shunt, additional mid muscular VSD, severe PAH	VSD shunt remained predominantly left to right. No increase in the pulmonary venous return	ASO + VSD closure	Mild TR with a gradient of 56 mmHg. Severe RV dysfunction
5	7 months	d-TGA, a large upper muscular VSD predominantly L-R shunt, additional mid-muscular VSD, small ASD, small PDA, severe PAH	VSD demonstrated a R-L shunt. Increased pulmonary venous return	ASO + VSD closure + ASD closure + PDA ligation	Mild TR with a gradient of 27 mmHg. Normal ventricular function
6	6 months	d-TGA, a large subarterial VSD predominantly L-R shunt, severe PAH	VSD demonstrated a R-L shunt. Increased pulmonary venous return	ASO + VSD closure	Trivial TR with a gradient of 20 mmHg. Good ventricular function

(IAS = intact atrial septum; IVS = intact ventricular septum; ASO = arterial switch operation; PAH = pulmonary arterial hypertension; DORV = double outlet right ventricle; ASD = atrial septal defect; VSD = ventricular septal defect; PDA = patent ductus arteriosus; APW = aortopulmonary window; TR = tricuspid regurgitation; RV = right ventricle; L-R = left to right; R-L = right to left)

based on changes in the echocardiography findings after oxygen administration. Four of those operable (i.e., four of five) patients underwent arterial switch operation and one patient underwent atrial switch surgery. The immediate surgical outcome was found satisfactory in all cases postoperatively, except in Case 2 who showed evidence of persistent moderate pulmonary arterial hypertension.

Although Case 4 still appeared inoperable on echo even after giving oxygen, the surgical team elected to proceed with an arterial switch operation due to his young age of 6 months. This patient demonstrated persistent PAH and severe RV dysfunction postoperatively.

Thus overall in five of six patients (83%), the postoxygen echo accurately predicted the outcome in patients with TGA-VSD physiology in whom operability was questionable.

DISCUSSION

Patients with TGA and a large post-tricuspid shunt are at risk for early onset and rapid development of severe pulmonary vascular obstructive disease (PVOD), which complicates the course of the patient.^[1] In fact, PVOD is a major determinant of both immediate and late surgical mortality after correction of the defect.

The evolution of arterial switch operation with the closure of the above-said defects in neonates and very young infants substantially reduces this hazard^[8] but unfortunately in our country many patients with this physiology go undetected for months, or sometimes years, and when they do reach the tertiary care center, either it is too late or it is too difficult to decide about the suitability for surgical correction. Therefore, in this subset of patients who present late to the hospital, surgery without information about the PVR always carries high morbidity and mortality. However, the status of the PVR and whether it is reversible or not is not always clear. The degree of PVOD can be directly assessed by lung biopsy,^[9] although this has distinct limitations and is not practical in our country's scenario.^[10,11]

The quantitative assessment of PVR by hemodynamic and saturation data obtained by cardiac catheterization does not always represent the true status of the patient. Unsure extent of mixing of two parallel circulations, especially in the presence of suspected pulmonary vascular disease in TGA-VSD physiology, always raises a doubt on pulmonary flow and PVR calculated by catheterization data. In addition, pulmonary arterial oxygen content is usually very high; the arteriovenous oxygen difference is therefore low, so that small variations in the measurement of oxygen saturations will result in major errors in the calculation of flow. The

application of the Fick principle tends to overestimate the pulmonary blood flow and hence underestimate PVR.^[12] Furthermore, bronchial artery hypertrophy has been shown angiographically^[13] and histologically^[14] suggestive of increased bronchial flow in TGA patients. If there is a large bronchial flow, the true oxygen saturation of precapillary blood would be considerably lower than that measured in pulmonary arteries. Therefore, the true pulmonary arterial oxygen saturation of blood perfusing the lungs would be lower and the flow calculated from the measured oxygen saturation would be higher than the actual flow.^[12,15] This error due to the effect of the bronchial circulation giving spuriously high estimates of pulmonary blood flow has been found to be greatest when oxygen consumption is low and aortic blood is very desaturated.^[16] In addition, in some cases, due to anatomical factors, the catheter is unable to enter the pulmonary arteries and then pulmonary flow and PVR cannot be obtained due to incomplete data. Additionally, catheterization itself is an invasive procedure and not free of side effects and complications especially in cyanotic patients.^[17] The fallacies of catheterization in such patients made us think about using oxygen administration followed by echo assessment to solve the issue of operability in older patients with TGA and a large post-tricuspid shunt.

It is well known that oxygen reduces pulmonary arterial pressure and PVR^[18,19] especially where irreversible PVOD has still not set in. This effect of oxygen is typically tested in the catheterization lab. Clinically, we attempted to analyze this effect of oxygen noninvasively in correlation with color Doppler echocardiography in patients with TGA-VSD physiology where operability was in question.

After giving oxygen, PVR fell in all our cases, except in Case 4, [Table 1] resulting in increased pulmonary blood flow as indicated by increased pulmonary venous return to LA. Additionally in Case 1, there was reversal of flow in the descending aorta in the absence of aortic regurgitation suggesting an increase in the left-to-right shunting of blood through the AP window which was not there prior to oxygen administration. In Cases 2, 3, 5, and 6, the shunt across VSD changed in the direction to right-to-left after oxygen administration, again suggestive of a drop in PVR.

An overall analysis shows that in five out of six study cases (i.e., 83%), postoxygen echo predicted operability accurately in patients with TGA-VSD physiology in whom operability assessment was not straightforward.

Although number of patients we studied were few, we found this method practically useful in determining the operability in TGA-VSD physiology. Description of any such method in this particular situation is not available in the literature.

CONCLUSION

Patients with TGA-VSD physiology who present late to the hospital, and in whom operability is doubtful on clinical assessment, can be subjected to the above-described method of determining operability using echocardiography after administering oxygen. Although not 100% accurate in predicting operability and postoperative pulmonary hypertension, this is a simple and noninvasive method that can aid in decision making in a tricky situation.

A large prospective study to assess the utility of this clinical tip needs to be performed.

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