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Pulmonary Atresia With an Intact Ventricular Septum: Preoperative Physiology, Imaging, and Management

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Abstract

Pulmonary atresia with intact ventricular septum (PA-IVS) is a rare complex cyanotic congenital heart disease with heterogeneous morphological variation. Prenatal diagnosis allows for developing a safe plan for delivery and postnatal management. While transthoracic echocardiography allows for detailed delineation of the cardiac anatomy, additional imaging modalities such as computed tomography, magnetic resonance imaging, and catheterization may be necessary to further outline features of the cardiac anatomy, specifically coronary artery anatomy. The size of the tricuspid valve and right ventricular cavity as well as the presence of right ventricle–dependent coronary circulation help to dichotomize between biventricular repair versus univentricular palliation or heart transplantation, as well as predicting the expected survival. The delineation and understanding of these features help to dictate both medical and surgical management.

Keywords

ductal stent, pulmonary atresia with intact ventricular septum, pulmonary valvotomy, right ventricle-dependent coronary circulation, systemic-to-pulmonary artery shunt

Introduction

Pulmonary atresia with intact ventricular septum (PA-IVS) is a rare complex cyanotic congenital heart disease with heterogeneous morphological variations.[1] The unique feature of this malformation is the lack of egress from the right ventricle (RV) into the pulmonary trunk along with varying degrees of tricuspid valve and RV hypoplasia. There are also associated abnormalities in the coronary circulation such as ventriculocoronary connections, fistulae, coronary stenosis, or atresia.[2] In approximately 9% to 34% of patients with PA-IVS there will be RV-dependent coronary circulation (RVDCC). The presence of coronary abnormalities, with or without RVDCC, can increase morbidity and mortality.[3-7]

In the setting of PA-IVS the pulmonary circulation is ductal dependent and this lesion often presents with cyanosis in the undiagnosed infant, and subsequently requires the establishment of a reliable means of pulmonary blood flow. The surgical approach (biventricular repair vs single ventricle palliation or heart transplantation), and resulting outcomes of these infants will mainly depend on the size of the tricuspid valve and RV, as well as the presence or absence of RVDCC.[6,8,9] In this review, we discuss the preoperative physiology, anatomical and functional assessment, and management in these patients.

Epidemiology

PA-IVS is a rare congenital malformation of the heart, accounting for approximately 3% of all congenital heart disease. It is present in approximately 4 to 8 per 100,000 live births and is the third most common form of cyanotic congenital heart disease.[10-12] The incidence of PA-IVS is undoubtedly greater in the fetal population as a subset of these patients, particularly those with severe tricuspid regurgitation, may succumb to fetal demise and others are electively terminated.[13] Males are more likely to have PA-IVS

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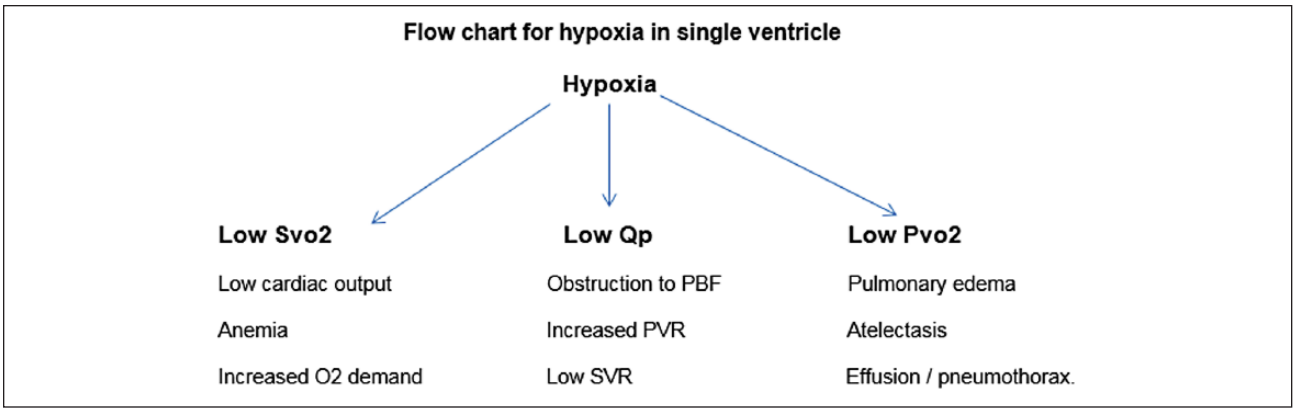

Figure 1. Flowchart for hypoxia in the single ventricle. O2, oxygen; PBF, pulmonary blood flow; Pvo2, pulmonary venous oxygen saturation; PVR, pulmonary vascular resistance; Qp, pulmonary blood flow; Svo2, systemic venous oxygen saturation; SVR, systemic vascular resistance

with a male to female ratio of 1.5:1. Also of note is that there is up to a 25% recurrence risk for CHD in siblings.[14]

Embryology

Kutsche and Van Mierop[15] postulated that PA-IVS is acquired later in gestation when compared with pulmonary atresia with a ventricular septal defect. They described that the pulmonary valve becomes atretic later in development following closure of the interventricular communication, a finding suggested by what is generally a well-developed pulmonary trunk. Further observations report that the angle of origin of the arterial duct from the aorta also correlates with the timing of development of pulmonary atresia. Those with pulmonary atresia with a VSD, or PA-IVS with a severely hypoplastic tricuspid valve and RV, will commonly have an arterial duct that inserts more proximal than normal on the aortic arch with an acute angle. This group also often includes those with ventriculocoronary connections. Comparatively, those with PA-IVS and a well-developed tricuspid valve and RV have a more normally positioned arterial duct that has a more obtuse angle of origin from the aorta.[15,16] With the advent of episcopic microscopy, the development, and hence maldevelopment, of the outflow tracts has now been further clarified. What was once described to develop from the "conus" and the "truncus," is now better described to derive from 3 components; the proximal (subvalvar outflow tracts), intermediate (arterial valves) and distal outflow tract (intrapericardial great arteries).[17] The precise morphogenesis of PA-IVS remains unclear, however. Some have reported evidence suggesting that atresia of the pulmonary valve is not the inciting insult but may actually be a result of abnormal coronary arterial development.[18]

Morphologic considerations aside, these patients commonly develop either tricuspid regurgitation or coronary fistulas, which help decompress a hypertensive RV. Additionally, the distinction between an imperforate valve (membranous atresia) and atresia of the entire outflow tract (muscular atresia) is important. Atresia of the outflow tract is usually associated with ventriculocoronary connections and coronary ostial stenosis, which portends a poorer outcome. Membranous atresia of the pulmonary valve, however, is not generally associated with coronary abnormalities and may allow for a less invasive approach such as percutaneous pulmonary valve perforation and balloon angioplasty.[19,20]

Hemodynamics and Physiology

In utero, the fetus with PA-IVS who develops severe tricuspid regurgitation can present with significant hydrops and intrauterine demise secondary to a low- pressure, severely hypoplastic, and failing RV. Those with a relatively normal sized tricuspid valve and RV often do not develop hydrops and commonly are born with stable hemodynamics.[21]

After birth, the arterial duct remains the sole source of pulmonary blood flow. Prenatal diagnosis allows for early initiation of prostaglandin E1 (PGE1) following birth to maintain the arterial duct as a stable source of pulmonary blood flow prior to surgical repair or palliation in the first few weeks of life. An atrial communication is necessary to permit the obligate right to left shunting of the systemic venous return, with complete mixing on the left side of the heart, resulting in expected cyanosis. In this functionally univentricular physiology with complete mixing of pulmonary venous and systemic venous blood, the distribution of systemic and pulmonary blood flow is dependent on the relative pulmonary and systemic resistances. As is seen in this setting, oxygen saturations in the aorta and pulmonary trunk is equal. The arterial saturation is therefore determined by three primary variables: the ratio of pulmonary to systemic blood flow (Qp: Qs), systemic venous saturation, and pulmonary venous saturation (Figure 1).

The subset of patients with a competent tricuspid valve and suprasystemic RV pressure are at increased risk of various coronary artery abnormalities such as ventriculocoronary connections and coronary ostial stenosis and atresia. These patients are also more likely to have RVDCC. In these patients, myocardial perfusion largely depends on perfusion pressure in the RV, and hence, they are at risk for myocardial ischemia and infarction, particularly if the high-pressure RV is decompressed.[22-24]

Imaging

Fetal Diagnosis

Standardization of outflow tract imaging, as outlined by the 2013 American Congress of Obstetricians and Gynecologists fetal imaging guidelines, has led to an improvement in fetal detection of various congenital malformations of the heart.[25,26] A complete fetal echocardiogram in patients suspected of having a congenital malformation of the heart is recommended at approximately 18 to 24 weeks' gestation, with standard guidelines for fetal echocardiogram described by the American Society of Echocardiography.[27]

Prenatal diagnosis of PA-IVS helps with risk stratification and selection of the most ideal palliative or corrective approach. Risk stratification and selection of a palliative or corrective approach is based on tricuspid valve morphology, RV morphology, the form of pulmonary valve atresia (membranous versus muscular), and the presence or absence of RVDCC.[11,28-30]

Because of these reasons, prenatal diagnosis aids in more patient-specific counseling. Specific fetal characteristics reported as being predictive for biventricular repair strategy include a tricuspid *Z*-score ≥−3, the absence of significant tricuspid regurgitation, absence of ventriculocoronary connections, and the absence of subaortic stenosis.[31,32] While RVDCC is clearly a predictor of poor outcomes, it is also the subset of patients with severe tricuspid regurgitation identified in utero, often associated with either Ebstein's anomaly of the tricuspid valve or tricuspid valve dysplasia, with resulting significant RV dilation and the so-called "wall-to-wall heart" appreciated on chest radiograph, which also hold an extremely poor prognosis. These patients may account for approximately one-sixth of those with PA-IVS.[33] Additional predictors include indirect assessment of the right atrial pressure using the tricuspid valve, as well as atrial communication and venous duct Doppler.[34] While prenatal diagnosis is certainly invaluable in assisting with early counseling for families, prenatal diagnosis has not been shown to decrease mortality rate.[21,29]

As previously mentioned, this functionally univentricular physiology requires an unrestrictive atrial communication to allow for egress of blood from the right heart to the left atrium for adequate systemic cardiac output. An oval foramen or deficiency in the floor of the oval fossa is universal in these patients, however, assessment of this communication in the fetus is prudent, as rarely this communication may need to be addressed in the cyanotic infant, and has even been addressed in fetal life.[35] More commonly, but also with limited experience to specialized, high-volume centers, fetal cardiac intervention is considered with pulmonary balloon valvuloplasty in those with moderate tricuspid and right ventricular hypoplasia and membranous PA-IVS.[36] Both of these fetal interventions have limited experience from non-randomized observatory studies with only short-term follow-up, making it difficult to draw conclusions at the present time.

Postnatal Preoperative Imaging

Transthoracic echocardiography is the mainstay to delineate the anatomy in PA-IVS; however, given the implications for various forms of coronary artery abnormalities, specifically when RVDCC is suspected, either cardiac catheterization or computed tomography is often necessary.

Atrial Septum. Detailed evaluation of the atrial septum to ensure adequacy of the atrial communication is crucial, although a patent oval foramen or deficiency in the floor of the oval fossa is universal in these patients. The direction of flow across the atrial communication should be demonstrated by color Doppler while the gradient across the communication should be quantified by spectral Doppler. Echocardiographic-guided balloon atrial septostomy may be necessary in the setting of a restrictive atrial communication[16]; however, this can usually be performed at the time of the initial surgical or interventional procedure rather than as an isolated emergent procedure. The atrial septum is best imaged in the subcostal long- and short-axis planes in the neonate. The size of atrial chambers should be documented, with the subset of patients with significant tricuspid regurgitation commonly having a dilated right atrium.

Tricuspid Valve. As mentioned, one of the major predictors of outcomes in PA-IVS is the size of the tricuspid valve. Measurements of valve annulus should be obtained in both the apical 4-chamber and parasternal long-axis views. Tricuspid valve *Z*-scores of approximately <−2 to −3 have been suggested as a cutoff to guide the surgical strategy toward univentricular repair (Figure 2).[21,37,38] Additionally, the morphology and function of the tricuspid valve apparatus must be delineated. Ebstein's anomaly has been described in near 10% of patients with PA-IVS, and will particularly be important to define and surgically address when biventricular repair is attempted.[16] The presence and degree of both tricuspid valve stenosis and regurgitation should be assessed by spectral and color Doppler (Figure 3). The tricuspid

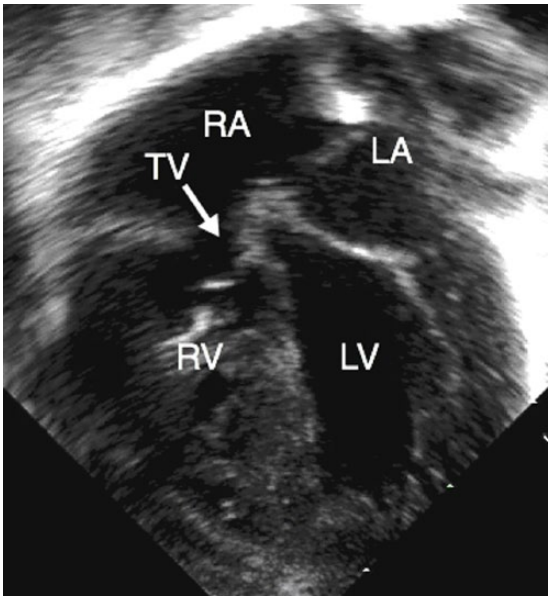

Figure 2. Apical 4-chamber echocardiographic view in an infant with pulmonary atresia and intact ventricular septum demonstrates a severely hypoplastic tricuspid valve (TV) and right ventricle (RV) with severe RV hypertrophy. LA, left atrium; LV, left ventricle; RA, right atrium.

regurgitation jet can estimate the RV pressure by continuous wave Doppler interrogation.

Right Ventricle. In the structurally normal heart, the RV is tripartite, consisting of an inlet, apical trabecular, and outlet components. In patients with PA-IVS, the RV remains tripartite in a literal sense; however, depending on the degree of RV mural hypertrophy, the cavity is "squeezed out", functionally creating either a tripartite, bipartite or unipartite ventricle (Figure 4).[39] This concept, that the RV in PA-IVS is always anatomically tripartite (in contrast to the incomplete RV present in double inlet right ventricle or tricuspid atresia), is important, as prediction of RV growth and function following surgical management for PA-IVS has been unpredictable. However, the RV and TV have both demonstrated the ability for significant growth following repair, which is largely because of the subsequent regression of mural hypertrophy.[40] Even so, it remains important to define the functional structure of the RV as this has been demonstrated to correlate with survival. A large, multicenter study demonstrated 74% 5-year survival in the functionally tripartite RV compared with 22% in the functionally unipartite RV.[16] The RV pressure is usually suprasystemic with various degrees of RV mural hypertrophy. The degree of hypertrophy is often related to the form of egress of blood from the RV which may either be in the form of significant tricuspid regurgitation or via ventriculocoronary connections. In the setting of significant tricuspid regurgitation, the RV tends to be thin-walled and dilated with a massively enlarged right atrium. In these instances, the RV may also demonstrate endocardial fibroelastosis. In contrast, the RV with ventriculocoronary connections tends to be severely hypertrophied and hypoplastic with a hypoplastic tricuspid valve. These features, along with assessment of RV systolic function, should be evaluated in all echocardiographic planes.[41,42]

Right Ventricular Outflow Tract and Pulmonary Arteries. Defining the type of pulmonary valve atresia (membranous versus muscular) is important for decisions on interventional or surgical management, with the former potentially allowing for initial intervention via catheterization in the absence of RVDCC. The right ventricular outflow tract can be evaluated in all planes, but is best evaluated in the subcostal and parasternal short-axis planes. The subtype of pulmonary atresia can be defined in both short and long axis parasternal planes (Figure 5). The branch pulmonary arteries are usually well developed, and should be defined in the parasternal short axis. Color Doppler interrogation across the RV outflow tract (RVOT) at low Nyquist limits will determine whether there is functional pulmonary atresia (suggested by the presence of pulmonary regurgitation) versus anatomical pulmonary atresia. The size and direction of blood flow through the patent arterial duct in this ductal-dependent lesion, which is expected to be exclusively left to right, can be defined in the high parasternal short axis, left parasternal ductal cut, and suprasternal notch views by both 2-dimensional imaging and color Doppler. Thorough delineation of the arterial duct is important, particularly for patients who are undergoing ductal stenting as a mode of establishing more reliable pulmonary circulation.[43]

Coronary Arteries. As previously mentioned, the coronary distribution is important for prognostication and decisions regarding management. Commonly encountered coronary artery abnormalities in PA-IVS include ventriculocoronary connections as well as coronary artery stenosis or atresia. In a subset of patients with PA/IVS, a large proportion of coronary perfusion may be retrograde from an RV with suprasytemic pressure. In this cohort RV decompression will likely be detrimental as decreasing the RV pressure will result in decreased coronary perfusion. RVDCC most often occurs when coronary stenosis or atresia occur proximal to ventriculocoronary connections. RVDCC has been described in anywhere from 3% to 34% of these patients with PA-IVS. This variability is not surprising as the definition of RVDCC varies between institutions.[5,44-47] Larger multicenter studies where a more uniform definition has been accepted have demonstrated a prevalence of 5% to 9%.[3,4,16] The Boston group has defined RVDCC as the

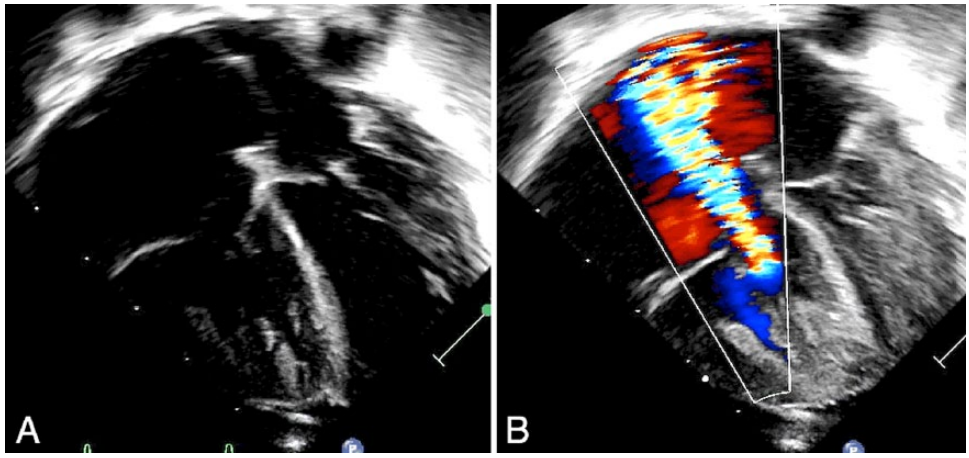

Figure 3. (A) Apical 4-chamber echocardiographic view in an infant with pulmonary atresia and intact ventricular septum demonstrates a dilated right atrium, with normal-sized tricuspid valve and right ventricle with mild right ventricular hypertrophy. There is poor coaptation of the tricuspid valve. (B) Color Doppler demonstrates moderate tricuspid regurgitation.

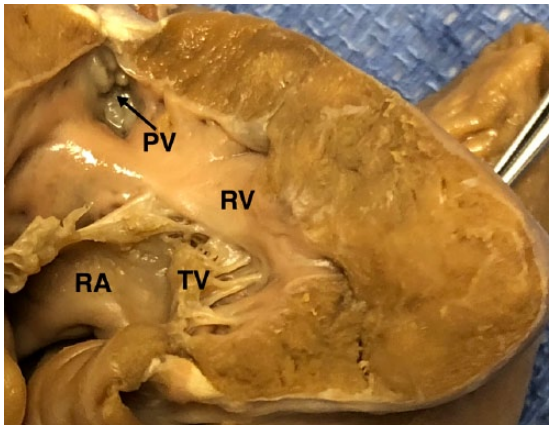

Figure 4. This specimen with pulmonary atresia and intact ventricular septum demonstrates severe hypertrophy of the right ventricle (RV). The RV remains tripartite, with an inflow portion guarded by a hypoplastic tricuspid valve (TV) and its supporting apparatus, the apical trabecular portion and the outflow with membranous pulmonary valve (PV) atresia. Because of the severe RV hypertrophy, however, the apical trabecular component is largely "squeezed out," creating a functionally bipartite RV. RA, right atrium.

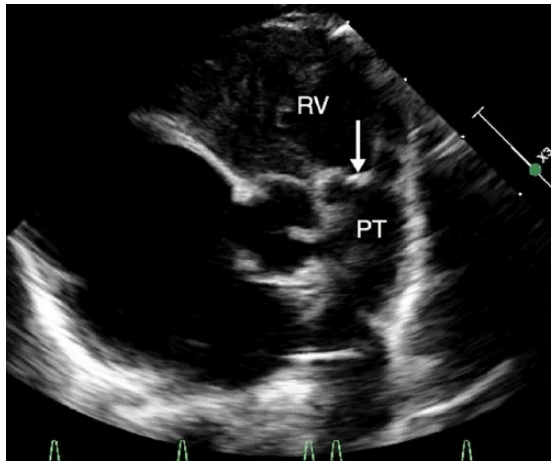

Figure 5. Parasternal short-axis echocardiographic view suggests membranous pulmonary atresia (arrow). This must be confirmed on color Doppler with a low Nyquist limit. PT, pulmonary trunk; RV, right ventricle.

presence of ventriculocoronary connections with angiographically severe obstruction of at least 2 major coronary arteries, complete aortocoronary atresia, or situations in which a significant portion of the left ventricular myocardium was supplied by the RV.[8] The proposed mechanism for adverse outcome after RV decompression in patients with RVDCC relates to diastolic runoff from the aorta through the coronaries into RV chamber, stealing from the coronary circulation and resulting in myocardial ischemia and infarction.[42,48]

The proximal coronary artery anatomy and the presence of ventriculocoronary connections can be delineated by echocardiography, with standard parasternal short- and long-axis views assessing the origins by both 2-dimensional imaging and color Doppler. Sweeps through the RV myocardium in multiple planes using a low Nyquist limit on color Doppler imaging should be performed to identify such connections. Bidirectional flow in the coronary arteries should also increase suspicion of ventriculocoronary connections (Figure 6).

If coronary artery abnormalities are suspected but not adequately delineated by echocardiography, cardiac catheterization with coronary angiography versus computed tomographic angiography is necessary (Figure 7).[48,49]

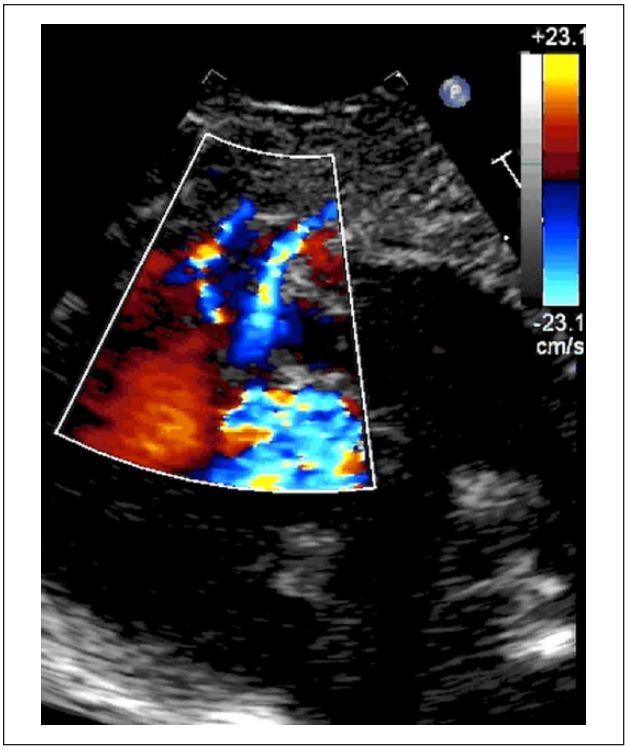

Figure 6. Parasternal short-axis color Doppler echocardiographic view in an infant with pulmonary atresia and intact ventricular septum at the level of the aortic root demonstrates reversal of flow in the right coronary artery and infundibular branch suggesting significant ventriculocoronary connections.

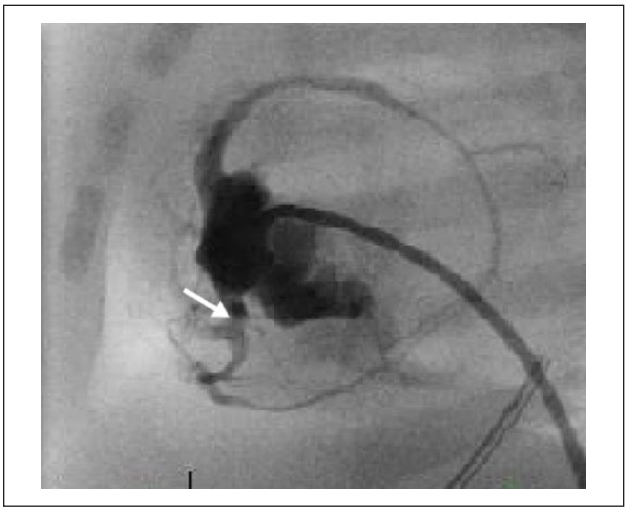

Figure 7. Lateral projection of a right ventriculogram in an infant with pulmonary atresia and intact ventricular septum demonstrating filling of the left and right coronary arterial systems via ventriculocoronary connections from the right ventricle (arrow).

Although cardiac catheterization is probably more commonly utilized, with improving technology excellent coronary artery delineation can be obtained by computed tomography angiography at radiation doses below that of standard diagnostic catheterization procedures and should be considered in this patient population.[50] Understanding the implications of these coronary abnormalities, a recent devised scoring system was proposed based on assessment of the amount of antegrade coronary perfusion to the named coronary arteries as evaluated by coronary angiography. This aortic perfusion score was found to be predictive, with good sensitivity and specificity, of need for heart transplantation or occurrence of death.[51] This concept supports the definitions of RVDCC as defined by the Boston group and its effects on outcomes, as well as the importance of thoroughly defining these abnormalities.[7,8]

Preoperative Management

Prompt initiation of prostaglandin therapy is crucial for survival in this ductal dependent lesion, and should be administered as soon as the diagnosis of PA-IVS is suspected in the postnatal period. Subsequent intubation and mechanical ventilation may be necessary because of apnea that may result from prostaglandin infusion. Neonates with PA-IVS should be transferred to a tertiary care center equipped for the management of patients with complex congenital heart disease. In the setting of functionally univentricular physiology, manipulation of the Qp:Qs, systemic venous saturations, and pulmonary venous saturations will dictate the resulting arterial saturations.

Interventional and Surgical Management

Once PA-IVS is diagnosed, the morphology of the tricuspid valve, RV, subtype of pulmonary valve, and the coronary arteries will help determine the interventional or surgical management strategy.[21,39,52] Surgical or interventional management is often employed within the first week of life. Based on the available evidence, we propose the general guidelines outlined in Figure 8.

Percutaneous Balloon Atrial Septostomy

There must be an obligate right to left shunt in the setting of unpalliated PA-IVS, which means that an unrestrictive atrial communication is necessary. Occasionally, the atrial communication will be restrictive, in which case percutaneous balloon atrial septostomy or surgical septectomy may be required as part of the initial palliative procedure. This is especially true if a systemic-to-pulmonary shunt will comprise the initial surgical management, with retention of functionally univentricular physiology. In fact, in a large multicenter study of patients with PA-IVS, in patients who underwent an initial systemic-to-pulmonary artery

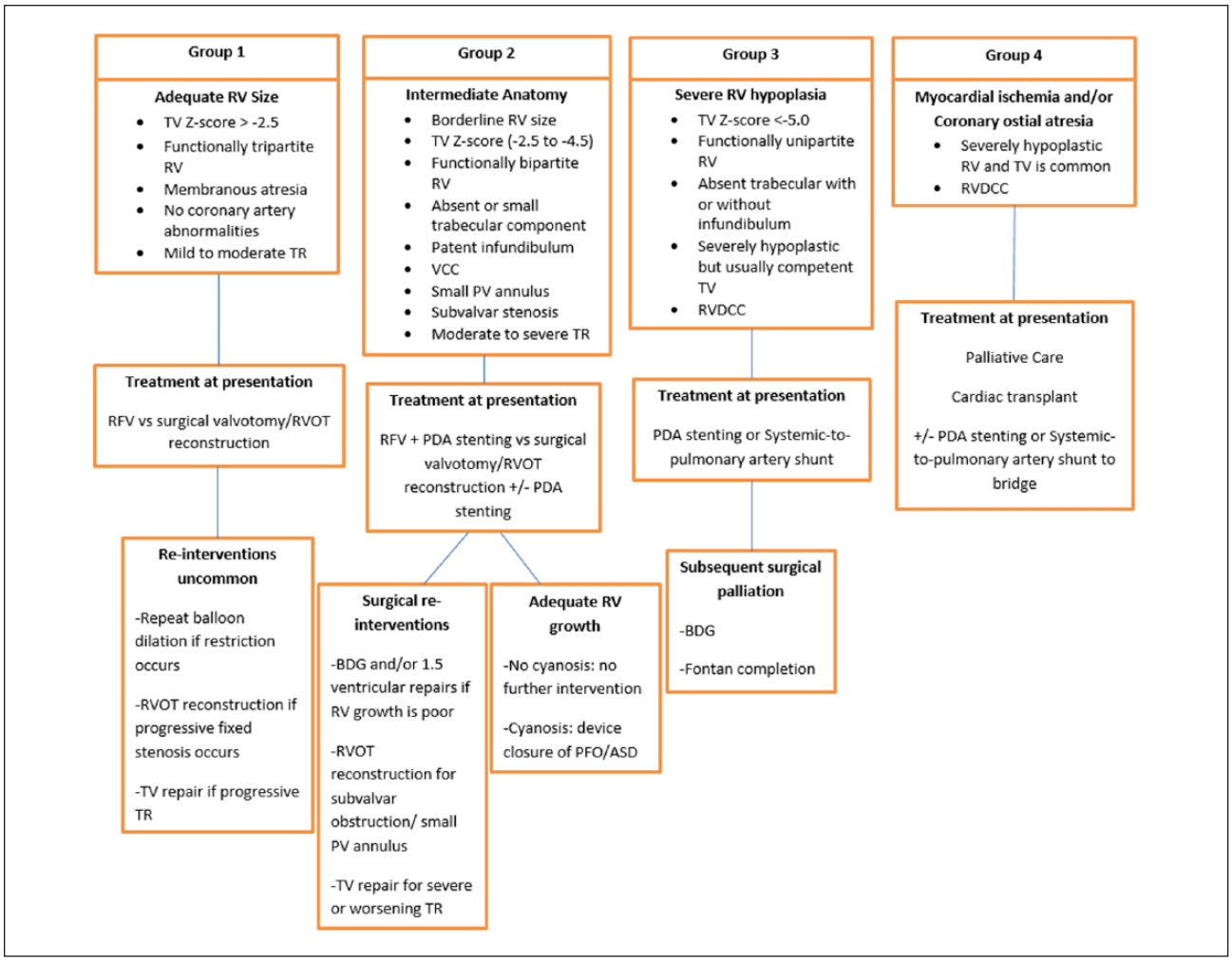

Figure 8. Suggested management strategies for morphological variants of pulmonary atresia and intact ventricular septum. ASD, atrial septal defect; BDG, bidirectional Glenn; PDA, patent arterial duct; PFO, patent foramen ovale; PV, pulmonary valve; RFV, radiofrequency valvotomy and balloon dilation; RV, right ventricle; RVDCC, right ventricle-dependent coronary circulation; RVOT, right ventricular outflow tract; TV, tricuspid valve; TR, tricuspid regurgitation; VCC, ventriculocoronary connections.

shunt without having had a balloon atrial septostomy performed earlier, approximately one-quarter of these patients eventually required a septostomy or septectomy performed at a later date because of concern for a restrictive atrial communication.[16] Although this procedure is rarely needed in an urgent fashion, it should be considered during the initial palliative procedure if the adequacy of the atrial communication is in question and the child will continue with functionally univentricular physiology.

Percutaneous Pulmonary Valvotomy

In membranous PA-IVS without RVDCC, transcatheter pulmonary valve perforation with a wire, laser, or radiofrequency energy, followed by balloon dilation of the valve, is a feasible option for initial palliation.[19,20,53] The arterial duct may or may not be stented at the time.

The establishment of biventricular circulation via transcatheter intervention requires the absence of RVDCC as well as an adequately sized tricuspid valve and RV to support pulmonary circulation. Such catheter-based intervention is not without risk. A review of 30 patients demonstrated a 17% prevalence of myocardial perforation. Additionally, 62% of patients required some form of subsequent surgical intervention to augment pulmonary blood flow in the same study. Thus, detailed anatomical assessment may lead to better patient selection and may improve interventional outcomes.[54]

Alwi et al[55] compared the outcomes of infants with PA-IVS undergoing radiofrequency ablation and balloon dilation versus surgical valvotomy and subsequent systemic-to-pulmonary artery shunt, reporting improved outcomes in the interventional group. In addition to an increased rate of complications and mortality, they also reported that patients who underwent an initial surgical

intervention more commonly required subsequent catheter-based interventions to further address the RVOT.[55] The study concluded that transcatheter intervention is an excellent option in patients with the appropriate anatomical substrate.

Surgical Repair Versus Palliation

When a surgical approach is deemed most appropriate, the previously discussed morphologic features will help decide between a biventricular and univentricular approach. Irrespective of the specific surgical strategy, the general goal is to establish a more definitive means of supplying pulmonary blood flow and to optimize the potential for growth of the RV and tricuspid valve.[56] General guidelines for surgical management are as follows.

The Dilated, Normal Sized, or Mildly Hypoplastic Right Ventricle

In patients with a mildly hypoplastic RV (the size of the RV is two-thirds of normal or greater with a tricuspid valve *Z*-score of 0 to −2) there is often a well-developed RVOT with membranous pulmonary atresia. In these patients, a surgical pulmonary valvotomy with RVOT reconstruction may be considered. The RV tends to be stiff and noncompliant, allowing these patients to do relatively well without a fully competent pulmonary valve. Placement of a systemic-to-pulmonary artery shunt to augment pulmonary blood flow, with ligation of the patent arterial duct, may also be appropriate if an additional source of pulmonary blood flow is felt to be needed. These patients may remain desaturated following surgical repair due to a noncompliant, hypertrophied RV although in the long-term most of these infants will tolerate a biventricular repair.[21,39,52,57,58]

The Severely Hypoplastic Right Ventricle

In the situation where there is a severely hypoplastic RV and tricuspid valve there will also often be ventriculocoronary connections. Functionally univentricular palliation is commonly warranted in these cases. Initial palliation includes a systemic-to-pulmonary artery shunt often in the form of a modified Blalock-Thomas-Taussig shunt, or patent arterial duct stenting, and is followed by the bidirectional Glenn and subsequent Fontan anastomosis. Additionally, patent arterial duct stenting or placement of a modified Blalock-Thomas-Taussig shunt can be used as an initial palliation strategy to bridge to heart transplant or destination therapy in a select patient population with PA-IVS such as those with RVDCC. Early mortality in those with RVDCC appears to be related to coronary ischemia at the time of initial palliation; however, functionally univentricular palliation in these patients yield excellent long-term results in the absence of coronary artery ostial atresia or frank myocardial ischemia.[8] Given the high mortality in PA-IVS with coronary artery ostial atresia, reported to be 100% in one high-volume institution's experience, many advocate for primary cardiac transplantation versus palliative care in this subset of patients.[8]

Postoperative Management

The major postoperative concerns relate to myocardial perfusion and subsequent ischemia, more commonly occurring after decompression of the RV in patients with RVDCC, and more specifically in those with coronary ostial atresia.[7] However, even patients without RVDCC are at risk for postoperative myocardial ischemia as revealed in an autopsy study around 3 decades ago, which revealed that approximately 50% of patients with PA-IVS without RV-to-coronary artery fistulas or coronary dysplasia died after either RV outflow reconstruction or placement of a systemic-to-pulmonary arterial shunt.[59] In those with RVDCC in whom RV decompression is undertaken, as the RV pressure falls, there is resulting "steal" of blood flow from the coronary arterial bed through these abnormal connections into the RV leading to impaired biventricular myocardial perfusion and myocardial ischemia.[24,60]

Long-Term Follow-Up

No matter the initial management strategy, these patients require lifelong follow-up by a cardiologist who specializes in the management of CHD. Although the survival rates of patients with PA-IVS have substantially increased with the majority of deaths occurring within the first postoperative year, and 15-year survival rates of all subtypes of PA-IVS patients being reported to be anywhere from 58% to 87%,[4,40] many patients living into adulthood may require re-intervention or succumb to late-onset arrhythmias.[61-63]

Conclusion

PA-IVS clearly comprises a heterogeneous group of patients, which requires accurate morphological and functional assessment to appropriately prognosticate and dichotomize into a biventricular repair versus univentricular palliative strategy. The preoperative management requires a thorough understanding of single ventricle physiology, including the necessity for unrestrictive atrial communication and a patent arterial duct. Both prenatal and postnatal echocardiography serve as the mainstay for accurate preoperative diagnosis and morphological assessment, with cross-sectional imaging, including coronary angiography, often being necessary to accurately define the commonly associated and clinically relevant coronary

artery abnormalities, including RVDCC. Understanding of these concepts has led to improved management decision making and outcomes in these patients.

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