CONGENITAL CARDIOLOGY TODAY
Timely News & Information for Congenital/Structural Cardiologists & Cardiothoracic Surgeons Worldwide
International Edition
Vol. 21 - Issue 2
February 2023

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Pulmonary Atresia with Intact Ventricular Septum – Part I

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Introduction

As stated in a recent paper in Congenital Cardiology Today,[1] review of other congenital heart defects (CHDs) will continue; in this article, we will discuss pulmonary atresia with intact ventricular septum. The senior author (PSR) has had interest in pulmonary atresia with intact ventricular septum over the years and published papers, reviews and book chapters on this subject.[2-8] Because of the large volume of material included, it is divided into two parts; these papers will provide an updated review of this lesion.

Pulmonary Atresia with Intact Ventricular Septum

Pulmonary atresia (PA) with intact ventricular septum (IVS) is a complicated CHD characterized by complete blockage of the pulmonary valve, two separate ventricles, and an open tricuspid valve (though it may be small and hypoplastic). It should be noted that this defect is completely different from PA with ventricular septal defect (VSD) which is within the tetralogy of Fallot spectrum regarding anatomic features, pathophysiologic derangement, management options, and outcomes. PA with IVS is an uncommon CHD and constitutes less than 1% of all CHDs and approximately 3% of critical CHDs presenting as neonates.[9] Treatment options for PA with IVS infants have evolved in the past five decades, as addressed elsewhere;[2-8] transcatheter intervention in the neonate appears to emerge as a desirable management choice in selected group of babies.

Pathologic Anatomy

Most patients with PA with IVS have normal right-to-left relationship of atria (atrial situs solitus). There is also normal ventricular localization with the morphologic right ventricle (RV) on the right and the morphologic left ventricle (LV) on the left. Atrioventricular concordance with the right atrium (RA) connected to the RV and the left atrium (LA) connected to the LV is present. Similarly, ventriculo-arterial concordance exists with the pulmonary artery arising from the RV and aorta (Ao) coming off of the LV; thus, the great vessels are normally related. Most cases of PA with IVS are seen in subjects with levocardia, i.e., normal left-sided heart. A diagrammatic representation of PA with IVS is illustrated in **Figure 1.**

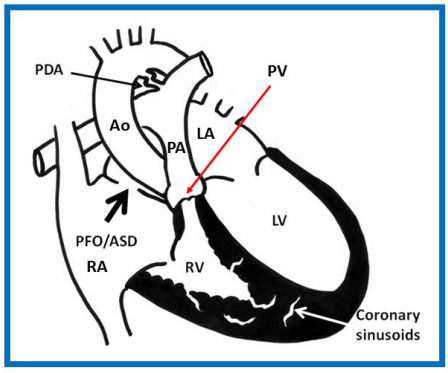

FIGURE 1 *Diagrammatic portrayal of pulmonary atresia with intact ventricular septum. The atretic pulmonary valve (PV) is shown with long red arrow. The thick arrow in the RA points to the patent foramen ovale/atrial septal defect (PFO/ASD) which is crucial to provide the egress of RA blood and avoid systemic venous obstruction. The RV cavity is small with thick musculature. Coronary sinusoids are shown in the RV myocardium. Patent ductus arteriosus (PDA) (small black arrow) is also marked and is essential to provide pulmonary blood flow. Right atrium (RA); left atrium (LA); right ventricle (RV); left ventricle (RV); aorta (Ao); pulmonary artery (PA). Modified from Reference 8.*

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Pulmonary Valve and Pulmonary Arteries

The pathology of the atretic pulmonary valve varies from one patient to the next. It may be near normal with well-formed pulmonary valve leaflets, but with commissural fusion in one extreme to infundibular atresia at the other end. Nonetheless, it is more often a membranous atresia of the pulmonary valve instead of muscular atresia.[10] Obviously, the pulmonary valve morphologic anatomy is pertinent if transcatheter relief of the obstruction is being considered.

The main and branch pulmonary arteries are usually well-formed and near normal in size; this is in distinct contrast to PA with VSD. In the latter the pulmonary arteries are usually hypoplastic, stenotic, or even discontinuous.

Right Ventricle

In the late 1950s, Greenwald, Davington and their associates[11,12] classified PA with IVS into two types: Type I, with a small or tiny RV cavity, and Type II with either a normal or a large RV cavity. But, sometime thereafter it was recognized that a much wider variation in the size of the RV cavity occurs, and several other classifications were introduced as reviewed in our prior publication.[3] Goor and Lillehei[13] introduced the concept of tripartite RV morphology in the mid-1970s. This concept was clinically applied by Bull, De Leval and their colleagues.[14,15] Normal RVs have three components: an inlet or sinus, a trabecular or apical component, and a conus or infundibulum. Bull and her associates[14] analyzed RV angiograms and pathologic specimens of patients with PA with IVS and identified three subsets: 1) Tripartite ventricles which exhibit all three components described above, 2) Bipartite ventricles with only inlet and infundibulum without trabecular cavity, and 3) Unipartite ventricles with only inlet and no trabecular and infundibular portions. de Leval and his associates[15] used the tripartite analysis to develop algorithms for surgical management of this defect. We have utilized these concepts in categorizing PA with IVS subjects:[7,8] 1) Tripartite RV consisting of all three components, namely, inlet, trabecular and outlet portions of near-normal size. This type of RV has the potential for subsequent RV enlargement, subject to adequate-sized tricuspid valve annulus, 2) Bipartite RV consisting of inlet and outlet components and absent trabecular portion. In this type, there is moderate hypoplasia of the RV. The adequacy of the RV growth is variable, and 3) Unipartite RV with only the inlet portion. The RV is markedly hypoplastic with high pressure and has the least chance for growth.

Tricuspid Valve

The tricuspid valve is typically small and occasionally near normal in size and at times large with regurgitation. The tricuspid valve Z scores appear to have relationship with morphology of the ventricle[14,15] as well the size of the RV.[16] Some patients may have a markedly stenotic tricuspid valve, mimicking tricuspid atresia.[17] Downward displacement of the tricuspid valve leaflets resulting in Ebstein's type of abnormality may be seen in <10% of cases. In addition, the tricuspid valve annulus diameter (Z score) helps to predict potential for future growth of the RV.[7,8,14-16]

Left Heart Structures

The left-sided structures, LA, LV and Ao, are usually normal. Enlargement of the LA and LV may be seen in subjects with large patent ductus arteriosus (PDA) and those who had large aorto-pulmonary shunts.

Inter-Circulatory Connections

Atrial Septum: Most commonly there is a patent foramen ovale (PFO) which allows obligatory right-to-left shunt **(Figure 1)**. Rarely, a true secundum atrial septal defect (ASD) may be seen.

Ventricular Septum: The ventricular septum is intact by definition; no VSD is seen.

Aorto-Pulmonary Connections: A PDA is present at birth which helps perfuse the lung **(Figure 1)**. But with time, there is a tendency for spontaneous closure with resultant pulmonary oligemia. Rarely, aorto-pulmonary collateral vessels may help to perfuse the lungs.

Myocardial Sinusoids

Myocardial sinusoids within the RV myocardium **(Figure 1)** may be present in a large number of patients; as many as 85% were seen in one study.[16] These sinusoids may be connected to the coronary arteries in nearly half of these patients. Such sinusoidal connections appear to be more common in subjects with small RV cavity and those with tricuspid valve Z scores less than –2.5. In nearly 10% of PA with IVS population,[5] there may be stenosis or atresia of the proximal coronary arteries. This will result in a situation where the distal coronary artery flow is dependent on high RV pressure; such a scenario is described as RV-dependent coronary circulation (RV-DCC). In patients with RV-DCC, reduction of RV pressure by surgical or transcatheter procedures is contra-indicated. Indeed, all diagnostic procedures in the neonate with PA with IVS should ensure absence of an RV-DCC before instituting procedures to relieve pulmonary valve obstruction.

Pathophysiology

Since the pulmonary valve is atretic, there is no forward flow from the RV to the pulmonary artery. Consequently, the RV blood regurgitates back into the RA. Then, an obligatory right-to-left shunt occurs across the atrial septum **(Figure 1)**. Consequently, systemic arterial desaturation is present in all patients with PA with IVS. Persistence of fetal circulatory pattern facilitates right-to-left shunt across the PFO. The pulmonary circulation is supplied by a PDA. However, there is tendency for spontaneous closure of the PDA with resultant pulmonary oligemia. Occasionally, the lungs are perfused via aortopulmonary collateral vessels. Some patients may have severe tricuspid regurgitation which may produce greater degree of RA dilatation, especially if the inter-atrial communication is restrictive.

The ease with which shunting across the PFO occurs and the degree of patency of the PDA determine the clinical features of babies with PA with IVS.

Clinical Features

The clinical features are largely determined by the patency of the ductus arteriosus and adequacy of PFO in decompressing the RA. At birth, the infants may appear normal and are asymptomatic. As the ductus begins to constrict, the infants will exhibit cyanosis and may have tachypnea and tachycardia. The symptomatology is directly related to the quantity of pulmonary blood flow which is proportional to the size of the ductus. Respiratory distress and metabolic acidosis will follow if the ductus is not opened with pharmacologic means. In subjects with restrictive PFO, systemic venous congestion develops with jugular venous congestion and

hepatomegaly; however, because of persistence of the fetal circulatory pattern, the PFO remains patent in most neonates. In addition, the proximal venous structures are compliant in the neonate and are less likely to exhibit signs of systemic venous congestion.

Physical examination shows cyanosis. Respiratory distress may also be present. A prominent left ventricular impulse is felt. The second heart sound is single. Sometimes a murmur of tricuspid insufficiency is heard best at the left lower sternal border. Even in patients with open ductus, it is unusual to auscultate a continuous murmur of PDA. In patients with obstructed PFO, hepatic enlargement with or without peripheral edema or ascites may be seen.

Differential diagnosis includes defects such as tetralogy of Fallot, tricuspid atresia, Ebstein's anomaly of the tricuspid valve, and critical pulmonary stenosis.[18-20] Findings in physical exam, chest x-ray and ECG are helpful in coming up with a diagnosis, but echocardiography is necessary for confirming the diagnosis.

Chest X-Ray

The chest roentgenogram **(Figure 2)** may demonstrate a normal sized heart or mild cardiomegaly with diminished pulmonary vascular markings. Cardiomegaly is more prominent in babies with severe tricuspid regurgitation, largely related to RA dilatation. In babies with wide-open PDA, pulmonary vascular markings are increased, although this may not be seen soon after birth because of high pulmonary vascular resistance. The main pulmonary artery segment may have concave appearance but may be covered by thymus in the early neonatal period.

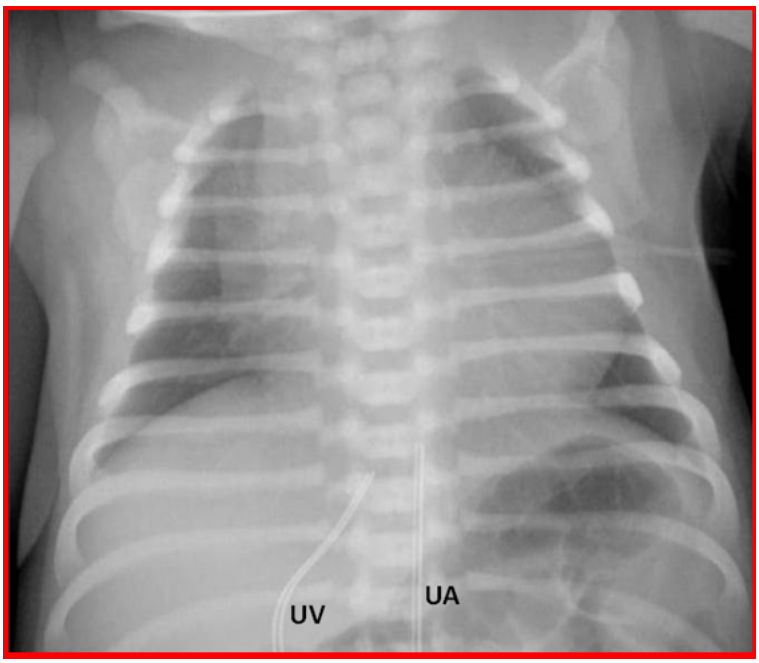

FIGURE 2 *Chest roentgenogram of a one-day old infant with pulmonary atresia with intact ventricular septum demonstrating moderate cardiomegaly, right atrial enlargement and decreased pulmonary vascular markings. Umbilical arterial (UA) and venous (UV) lines are labeled. Reproduced from Reference 8.*

Electrocardiogram

Electrocardiogram (ECG) shows a QRS axis between 0 and +90^0, enlargement of the RA, and left ventricular predominance or hypertrophy **(Figure 3)**. The RV electrical forces are minimal or low in most patients. However, some patients may show signs of RV hypertrophy, particularly in patients with tripartite ventricles. Patients with subendocardial ischemia, either due to high pressure in RV cavity or related to RV-DCC, may exhibit ST segment depression with or without T wave inversion. Ischemia from coronary sinusoids is not typically seen in the neonate.

In cyanotic babies with decreased pulmonary blood flow, ECG is very helpful in the differential diagnosis **(Figure 4)**. The usual causes are tetralogy of Fallot, pulmonary atresia or stenosis with intact ventricular septum, tricuspid atresia, and complex forms of pulmonary stenosis.[20,21] Babies with tetralogy of Fallot will show right axis deviation (+90 to ±180°) and RV hypertrophy. Neonates with PA with IVS, as detailed above will have an axis of 0 to +90°. Babies with tricuspid atresia demonstrate left axis (superior vector) deviation (0 to –90°). They also have LV hypertrophy and decreased right-ventricular voltages. However, babies with complex forms of pulmonary stenosis have variable findings.[20,21]

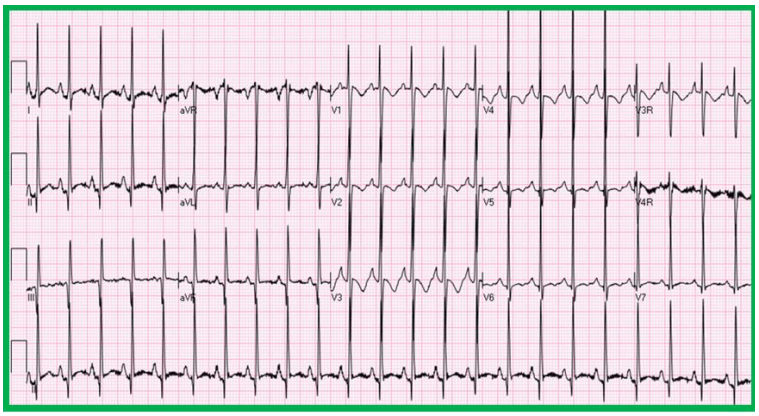

FIGURE 3 *Electrocardiogram of the same baby in Figure 2 shows +40^0 frontal plane axis, right atrial enlargement and left ventricular dominance. Reproduced from Reference 8.*

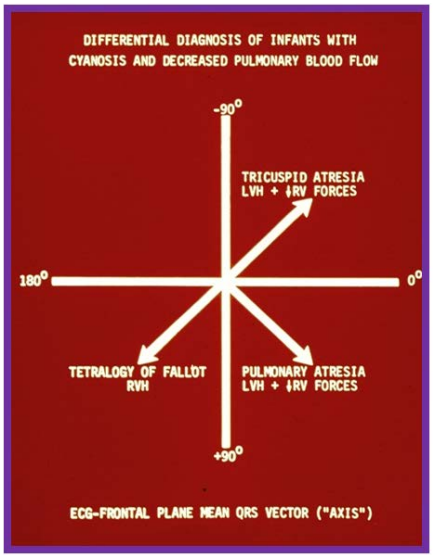

FIGURE 4 *This diagram illustrates the usefulness of the frontal plane mean QRS vector (axis) in assisting differential diagnoses of cyanotic heart defects with reduced pulmonary blood flow. A frontal plane axis between 0 and –90° is indicative of tricuspid atresia, an axis between 0 and +90° is suggestive of pulmonary atresia with intact ventricular septum while an axis between +90° and ±180° is seen with tetralogy of Fallot. The associated ECG abnormalities are also shown in the appropriate quadrant. Cyanotic babies with complex forms of severe pulmonary stenosis/atresia may have an axis in any quadrant. LVH, left ventricular hypertrophy; RV, right ventricle; RVH, right ventricular hypertrophy. Modified from Reference 20.*

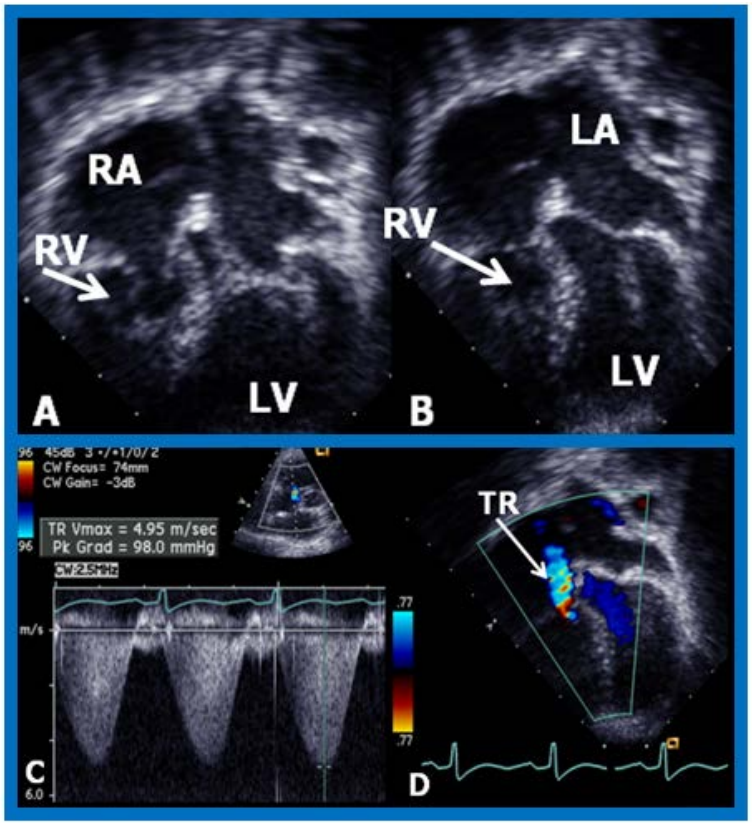

FIGURE 5 *Selected images from two-dimensional echocardiogram in apical four chamber views illustrating a very small/hypoplastic right ventricle (RV) with open **(A)** and closed **(B)** tricuspid and mitral valve leaflets (not labeled). The right atrium (RA) is dilated. **C** and **D**. Continuous wave **(C)**, and color **(D)** Doppler images demonstrating tricuspid regurgitation (TR). High TR Doppler velocity **(C)** is suggestive of high RV pressure. Left atrium (LA); left ventricle (LV). Modified from Reference 8.*

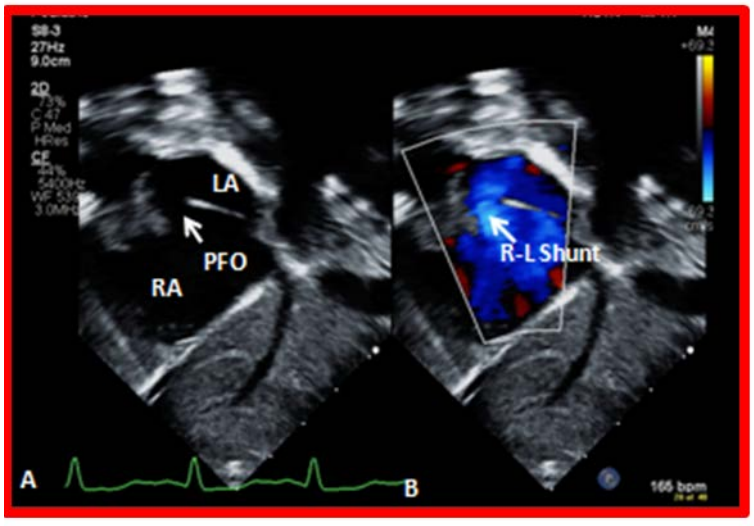

FIGURE 6 *Selected video frames from a subcostal echocardiographic study demonstrating patent foramen ovale (PFO) (arrow in **A**) with right to left shunt (R-L Shunt) across the PFO (arrow in **B**). Note laminar flow across the PFO in B. Left atrium (LA); right atrium (RA). Reproduced from Reference 26.*

Echocardiogram

Echocardiography is useful in confirming the diagnosis of PA with IVS and in defining chief issues relevant in management of PA with IVS.[22] Two-dimensional echocardiography (2D) will show an atretic pulmonary valve, small right ventricle, small but patent tricuspid valve, and tricuspid insufficiency **(Figure 5)**. Once the diagnosis of PA with IVS is established, other items, namely, Z scores of the tricuspid valve, right ventricular size and morphology, pulmonary valve morphology and annular diameter, size and patency of the pulmonary arteries, and shunting across the PFO/ASD and PDA should be evaluated. Finally, imaging of the RV myocardium to detect coronary sinusoids and to ensure lack of RV-DCC should be undertaken.

Z Scores of the Tricuspid Valve

The diameter of the tricuspid valve annulus should be measured in frozen video frames and normalized to body surface area, thus deriving Z scores. The tricuspid valve Z scores are generally proportionate to the size of the RV and have potential in predicting the RV growth.[7,8] Equally important is that Z-scores less than –3 are associated with the presence of coronary sinusoids and perhaps the presence of an RV-DCC.[23] In addition, tricuspid valve Z-scores better than –3 are seen with adequate RV growth and successful biventricular repair.[7,8,23]

Tricuspid to Mitral Valve Annulus Ratio

Ratio of tricuspid to mitral valve annulus should also be measured in frozen video frames and are useful in predicting the RV growth; ratios > 0.7 have better probability for RV growth while ratios < 0.5 are associated with lack of RV growth.

Right Ventricular Size and Morphology

The size and morphology of the RV may be defined by echo studies; the RV is usually small with hypertrophied RV musculature **(Figure 5)**. Further definition of RV morphology, classifying it into tripartite, bipartite or unipartite RV should be made: tripartite when all three components, namely, inlet, trabecular and outlet are present; bipartite with inlet and outlet portions are identified; and unipartite with only inlet cavity. These can be seen by reviewing parasternal and subcostal sagittal views imaging the RV. The tripartite and bipartite RVs may be addressed with either transcatheter or surgical relief of the atresia. By contrast, the unipartite RVs may have to be treated by univentricular palliation.[7,8,14] In addition, unipartite RVs may have coronary sinusoids and RV-DCC.[24,25]

Pulmonary Valve Morphology and Annular Size

The diameter of pulmonary valve annulus and the thickness and mobility of the valve leaflets can be defined by echo which may help in selecting treatment options.[7,8] On occasion, there may be difficulty in distinguishing between severe pulmonary valve stenosis and pulmonary valve atresia; if pulmonary insufficiency jet is visualized by color Doppler, it is likely to be pulmonary valve stenosis.

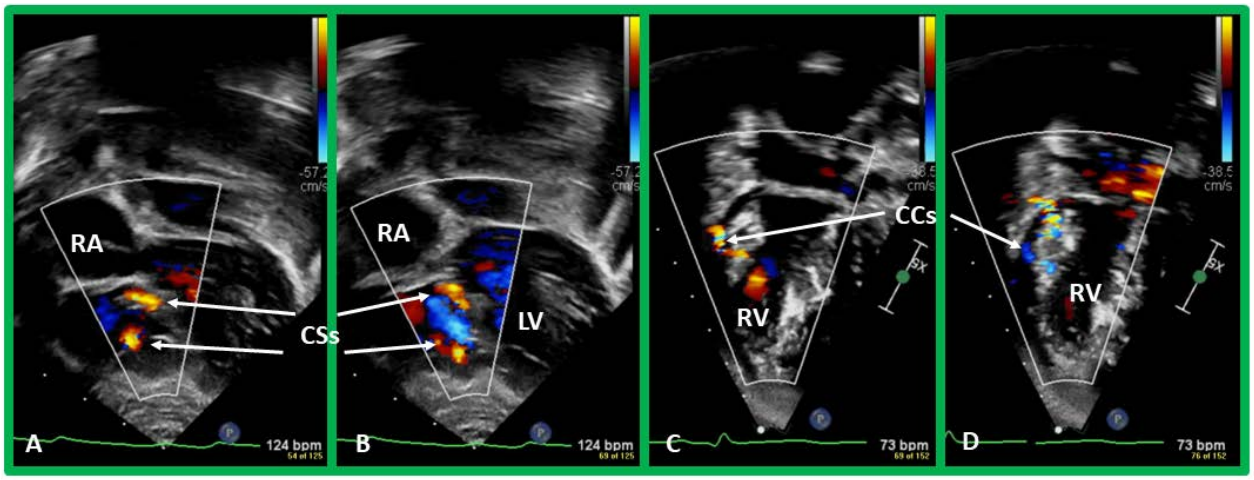

FIGURE 7
*Selected video frames from subcostal echocardiographic studies demonstrating coronary sinusoids (CSs) in **A** and **B** and coronary connections (CCs) in **C** and **D**. Left ventricle (LV); right atrium (RA); right ventricle (RV).*

Shunting Across the PFO/ASD

As discussed previously, an obligatory right-to-left shunt occurs across PFO/ASD. The PFO usually remains patent because of persistence of fetal circulatory pattern, and it is unusual to see obstruction during the neonatal period **(Figure 6)**. Beyond the neonatal period, there is a possibility for restriction at the PFO level. A small opening on subcostal 2D imaging, especially if it is associated with turbulent flow or a high Doppler velocity, is indicative of obstruction at the PFO.

Patent Ductus Arteriosus

Since there is pulmonary atresia, the pulmonary circulation is completely dependent upon the patency of the ductus. The PDA is open at birth, but tends to undergo spontaneous closure with time. The patency of the ductus can easily be demonstrated by color Doppler study. The PDA frequently originates from the usual location, but sometimes it may come off of the undersurface of the aortic arch. It is usually tortuous. On rare occasions, the pulmonary circulation is supported by aortopulmonary collateral vessels (which is more frequent in patients with PA with VSD).

Coronary Sinusoids and RV-DCC

The RV myocardium should be color flow Doppler interrogated to detect any coronary sinusoids and their connection to the coronary arteries **(Figure 7)**. While the proximal coronary arteries can be imaged,[27,28] atresia or narrowing of more distal coronary arteries may be more difficult to document by echo imaging.[22] Other imaging studies including aortic root or selective coronary angiography may be necessary to accomplish this goal. While RV-DDC may be suspected on echo studies, angiography may be necessary to confirm RV-DCC.

Cardiac Catheterization and Selective Cineangiography

Cardiac catheterization with selective cineangiography, while not needed for diagnosis of PA with IVS, it may be necessary to confirm/exclude coronary artery stenosis/interruption in subjects suspected of having large coronary sinusoids with coronary connections and RV-DCC. It is also useful in precise delineation of RV size and morphology,

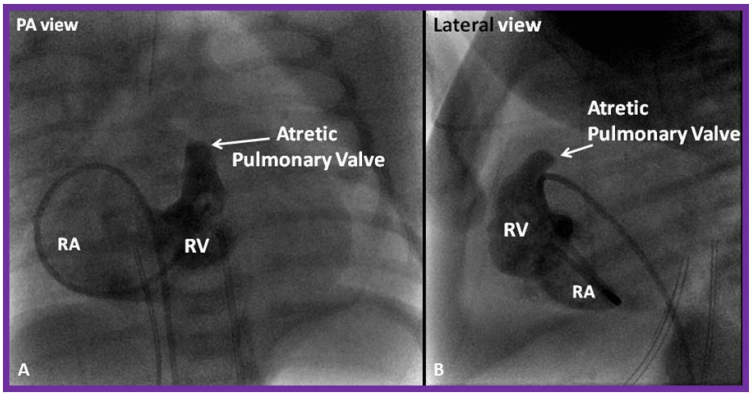

FIGURE 8 *Selected cine frames from a right ventricular (RV) angiogram in postero-anterior (PA) **(A)** and lateral **(B)** views in a neonate demonstrating a moderately hypoplastic tripartite RV. Tricuspid regurgitation opacifying the right atrium (RA) is also seen. The RV outflow tract ends blindly at the atretic pulmonary valve (arrows). Reproduced from Reference 8.*

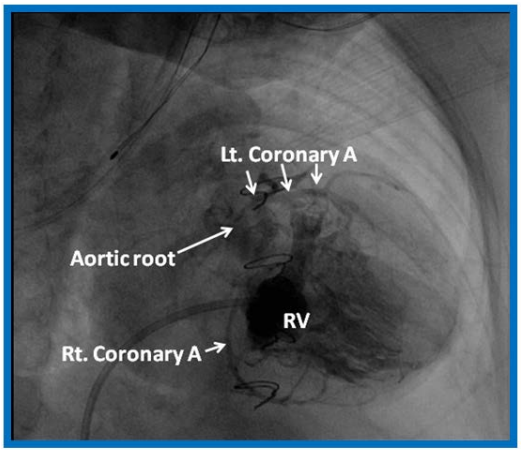

FIGURE 9 *Right ventricular (RV) cine-angiogram in right anterior oblique view demonstrating a smallish RV with subsequent opacification of both the right (Rt.) and left (Lt.) coronary arteries (A). The retrograde coronary flow exits into the aortic root. There is no evidence for coronary artery stenosis. Given these findings, this patient does not have an RV-dependent coronary circuit. Reproduced from Reference 8.*

preparatory to catheter-based procedures to establish RV to PA connection. The aims of the procedure are to define the size and morphology of the RV **(Figure 8)** and to exclude atresia or stenosis of proximal coronary arteries by aortic root or LV angiography, and if necessary, by selective coronary angiography in an attempt to exclude RV-DCC. Sometimes, RV angiography may opacify the sinusoids with retrograde opacification of coronary arteries **(Figure 9)**.

Summary and Conclusions

PA with IVS is a rare complex CHD and constitutes less than 1% of all CHDs. It is characterized by atresia of the pulmonary valve and variable degrees of hypoplasia of the RV. Important associated abnormalities such as hypoplasia of the tricuspid valve, PFO and RV-DCC are also present. Clinical, roentgenographic and electrocardiographic features are useful, but not confirmatory in diagnosing this entity; however, echo-Doppler studies and angiography are useful in assessing the anatomy and pathophysiology of this defect. It may be concluded that PA with IVS can be successfully assessed with the presently available diagnostic methods.

References

1. Rao PS. Congenital corrected transposition of the great arteries. Congenital Cardiology Today. 2022; 20(8):1-8.
2. Rao PS, Liebman J, Borkat G. Right ventricular growth in a case of pulmonic stenosis with intact ventricular septum and hypoplastic right ventricle. Circulation 1976; 53:389-394.
3. Rao PS. Comprehensive management of pulmonary atresia with intact ventricular septum. Ann Thorac Surg 1985; 40:409-413. doi: 10.1016/s0003-4975(10)60082-3.
4. Siblini G, Rao PS, Singh GK, Tinker K, Balfour IC. Transcatheter management of neonates with pulmonary atresia and intact ventricular septum. Cathet Cardiovasc Diagn 1997; 42:395-402.
5. Rao PS. Pulmonary atresia with intact ventricular septum. Current Treatment Options in Cardiovasc Med 2002; 4:321-336. doi: 10.1007/s11936-002-0012-6.PMID: 12093389.
6. Rao PS. Current management of pulmonary stenosis and atresia with intact ventricular septum. In: Heart Disease in Neonates and Children, Al Fagih, MR (ed). The Medicine Group, Oxford, England. 1985:109-120.
7. Balaguru D, Rao PS. Pulmonary atresia with intact ventricular septum. In. Rao PS, Vidyasagar D. (editors). Perinatal Cardiology: A Multidisciplinary Approach, Minneapolis, MN, Cardiotext Publishing, 2015.
8. Balaguru D, Rao PS. Pulmonary atresia with intact ventricular septum. In. Rao PS, Vidyasagar D. (editors), A Multidisciplinary Approach to Perinatal Cardiology, Volume 2. Cambridge Scholars Publishing, New Castle upon Tyne, UK. 2021:380-403. ISBN-13: 978-1-5275-6744-3; ISBN-10: 1-5275-6744-3.
9. Ferencz C, Rubin JD, McCarter RJ, et al. Congenital heart disease: Prevalence at live birth. The Baltimore-Washington Infant Study. Am J Epidemiol 1985; 121:31-36.
10. Kutsche LM, Van Mierop LH. Pulmonary atresia with and without ventricular septal defect: A different etiology and pathogenesis for the atresia in the 2 types? Am J Cardiol 1983; 51:932-935.
11. Greenwald WE, Dushane JW, Burchell HB, et al. Congenital pulmonary atresia with intact ventricular septum: two anatomic types [abstract]. Circulation 1956; 14:945-951.
12. Davington AL, Greenwald WE, Dushane JW, et al. Congenital pulmonary atresia with intact ventricular septum: clinico-pathologic correlation of two anatomic types. Am Heart J 1961; 62:591-602.
13. Goor DA, Lillehei CW, eds: Congenital Malformations of the Heart. New York: Grune & Stratton; 1975.
14. Bull C, de Leval MR, Mercanti C, et al. Pulmonary atresia with intact ventricular septum: a revised classification. Circulation 1982; 66:266-272.
15. De Leval MR, Bull C, Stark J, et al. Pulmonary atresia with intact ventricular septum: surgical management based on revised classification. Circulation 1982; 66:272-280.
16. Hanley FL, Sade RM, Blackstone EH, et al. Outcomes in neonatal pulmonary atresia with intact ventricular septum. J Thorac Cardiovasc Surg 1993; 105:406-427.
17. Rao PS. Is the term "tricuspid atresia" appropriate? Am J Cardiol 1990; 6:1251-1254.
18. Perloff JK. Pulmonary atresia with intact ventricular septum. In. The Clinical Recognition of Congenital Heart Disease. WB Saunders. Philadelphia 1978:429-438.
19. Bialostozky D, Attie F, Lupi E, et al. Atresia pulmonary con septum interventricular intacto. Arch Inst Cardiol Mex 1974; 44:195-210.
20. Rao PS. An approach to the diagnosis of cyanotic neonate for the primary care provider. Neonatology Today 2007; 2 (6):1-7.
21. Rao PS. An approach to the diagnosis of cyanotic neonate for the primary care provider. In. Rao PS, Vidyasagar D. (editors), A Multidisciplinary Approach to Perinatal Cardiology, Volume 1. Cambridge Scholars Publishing, New Castle upon Tyne, UK. 2021:295-312. ISBN-13: 978-1-5275-6744-3; ISBN-10: 1-5275-6744-3.
22. Freedom RM. Pulmonary atresia with intact ventricular septum. In. Pediatric Cardiovascular Medicine. Edited by Moller JH, Hoffman JIE. New York: Churchill Livingstone: 2000:442–460.
23. Satou GM, Perry SB, Gauvreau K, Geva T. Echocardiographic predictors of coronary artery pathology in pulmonary atresia with intact ventricular septum. Am J Cardiol 2000; 85:1319-1324.
24. Cheatham J. The transcatheter management of the neonate and infant with pulmonary atresia and intact ventricular septum. J Intervent Cardiol 1998; 11:363-387.
25. Ahmed AA, Snodgrass BT, Kaine S. Pulmonary atresia with intact ventricular septum and right ventricular dependent coronary circulation through the "vessels of Wearn". Cardiovasc Pathol 2013; 22:298-302.
26. Rao, P.S. Echocardiography in the diagnosis and management of tricuspid atresia. Appl. Sci. 2021, 11, 9472. https://doi.org/10.3390/app11209472.
27. Jureidini SB, Marino CJ, Rao PS. Congenital coronary artery anomalies. Indian J Pediatr 1998: 65:217-229.
28. Jureidini SJ, Marino C, Rao PS, et al. Transthoracic color and pulsed Doppler echocardiographic assessment of normal coronary artery flow in children. J Am Soc Echocardiogr 1998: 11:409-420.

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The Lead Skirts – Pregnancy in the Cath Lab

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I consider myself luckier than most. The first woman interventionalist I met was on the categorical cardiology fellowship interview trail. She had four children, and was living, breathing proof that a woman could be a cath doctor and mom – at the same time! During my cardiology fellowship, the 4th year interventional fellow we trained was a woman. I later went on to have a woman Cath Lab Director during my interventional 4th year, and a woman Cath Lab Director where I accepted my first job. I therefore was fortunate enough to have multiple women I could turn to for mentorship and advice on how to navigate the gender-specific concerns associated with a career as an interventional cardiologist. I later came to understand how unique my situation was, as I met other women interventionalists who found themselves essentially alone in this regard. It was through these interactions that I decided to organize the first Women in Pediatric Interventional Cardiology Symposium in 2018. This served as an opportunity for women in the field to meet and network, from which PbS, 'The Lead Skirts,' was born. An online organization, this group is a forum for women at various stages in their career, to interact and discuss topics specific to women pediatric interventionalists. With this column, we plan to introduce some of these gender-specific topics to the general congenital cardiology community in hopes of generating thoughtful discussion and collaborative efforts to achieve a better understanding and gender equity.
Sara Trucco, MD

Pregnancy in the Cath Lab

In the Unites States, federal law prohibits discrimination against pregnant workers. Despite this, a survey conducted in 2011 revealed that as many as 35% of respondents worked in a practice or hospital prohibiting women from continuing to work in the catheterization laboratory while pregnant.[1] Legally, fetal radiation dose accumulates only after the pregnancy is declared. Some women, therefore, are faced with the decision of being forced to not work in their chosen field, or continuing to work without proper fetal monitoring – neither of which is acceptable. This article aims to provide a review of available data regarding radiation exposure risk to pregnant workers, followed by individual pregnancy experiences reported by members of The Lead Skirts.

Radiation exposure, and its potentially harmful effects on a developing fetus, are often quoted as a significant factor preventing women from entering the field of interventional cardiology. A growing body of knowledge suggests the radiation risks associated with pregnancy in the catheterization laboratory can be safely mitigated with proper monitoring and adequate radiation reduction measures in place.[1,2] It is estimated that background radiation exposure averages 0.75 to 1 mSv per year, with variation based on geographic location. According to National Council on Radiation Protection (NCRP) estimates, exposure levels above 10 mSv may increase the risk of a child being born with a congenital malformation or cancer by 0.1% (**Table 1**).[3,4] In comparison, the typical interventional cardiologist is exposed to 1 to 4 mSv/year, though this range may not reflect the reduced radiation exposure rates achievable with current fluoroscopy systems and radiation reduction protocols.[2,5,6] The fetus is most vulnerable to radiation during the first trimester, while exposure later in gestation is associated with growth restriction and intellectual limitation (**Table 2**).[7,8] The threshold for these effects, however, is much higher than one would expect to receive under a protective lead apron, and is more than 10 times the safe limits recommended by the American College of Obstetrics and Gynecologists (ACOG) and the NCRP.[2,7,9] The Society of Cardiovascular Angiography and Interventions (SCAI) therefore, issued a consensus statement concluding, "Current data do not suggest a significant increased risk to the fetus of pregnant women in the cardiac catheterization laboratory and thus do not justify precluding pregnant physicians from performing procedures in the cardiac catheterization laboratory." It is essential, however, that radiation exposure during pregnancy be monitored closely with an additional fetal badge, as early as possible, and that properly fitting protective garments of at least 0.5 mm lead equivalent are worn throughout the pregnancy.

The decision to work in the catheterization laboratory while pregnant is multifactorial, and radiation exposure is only one aspect to consider. The physical strain of standing in lead for multiple procedures can cause musculoskeletal strain, more so in pregnancy when ligaments and tendons loosen. Additionally, the potential impact the physical and emotional stress of working in the catheterization lab may have on the development of hypertension, premature labor, or other pregnancy complications, is not well understood. Ultimately, each individual woman must consider these factors along with those specific to her family situation and career plans. It is imperative, however, that women be permitted to make this choice and do so in an environment that is respectful and supportive of this decision.
Sara Trucco, MD

TABLE 1 *Probability of Birth Without Malformation or Childhood Cancer*

Conceptus dose above back-ground, mSv	No malformation, %	No childhood cancer, %	No malformation and no child-hood cancer, %
0	96	99.93	95.93
0.5	95.999	99.926	95.928
5	95.99	99.89	95.88
10	95.98	99.84	95.83

This table is referenced from Circ Cardiovasc Interv. 2021;14:e009636. DOI: 10.1161/CIRCINTERVENTIONS.120.009636.

Experience and Perspectives from Women in Pediatric Interventional Cardiology

For many reasons, I decided to defer pregnancy until after I completed my interventional training. After completing my 4th year, however, I wasted no time and showed up to my first job early in my first trimester. I was new to the institution, and when to reveal my pregnancy to my new colleagues and staff weighed heavily on my mind. I was able to confidentially obtain a fetal badge, and eventually announced my pregnancy when genetic screening came back normal. I thankfully felt wonderful while pregnant, and found that if I ate regularly, I had no nausea or vasovagal symptoms. My partner was exceptionally supportive,

TABLE 2 *Effects of Gestational Age and Radiation Dose on Radiation-Induced Teratogenesis*

	Effects	Estimated threshold dose*
Gestational period		
Before implantation (0–2 wk after fertilization)	Death of embryo or no consequence	50–100 mGy
Organogenesis (2–8 wk after fertilization)	Congenital anomalies (skeleton, eyes, and genitals)	200 mGy
	Growth restriction	200–250 mGy
Fetal period		
8–15 wk	Severe intellectual disability (high risk)	60–310 mGy
	Intellectual deficit	25 IQ point loss/1000 mGy
	Microcephaly	200 mGy
16–25 wk	Severe intellectual disability (low risk)	250–280 mGy

This table is referenced from Circ Cardiovasc Interv. 2021;14:e009636. DOI: 10.1161/CIRCINTERVENTIONS.120.009636.

despite the fact she would later be left to cover cath call for the entirety of my maternity leave. My cath lab staff went out of their way to help in numerous ways: ensuring my radiation exposure was as low as possible, encouraging me to unscrub and remove my lead as soon as the case was completed, and providing what seemed like an endless supply of snacks! I wore double lead, even though the radiation safety officer at my hospital told me it wasn't necessary. Doing so gave me peace of mind, and my fetal dosimeter badge consistently reported "zero" or "indetectable" radiation levels. Once I reached full term, I was taken off the cath schedule given the unpredictability of when I would deliver. Instead, I worked in our outpatient clinic, seeing my regular clinic panel in addition to staffing the "72-hour access clinic" designed for new patients seeking timely outpatient cardiology consultation.

My first son was born at 39 weeks to the day, without complication. Having been on faculty less than a year, I was only eligible for six weeks of maternity leave. I used some vacation time and returned to work when my son was eight weeks old. It was then that the real work began! Finding the place and time to pump between cases, eat, prep the next case, while writing orders on the prior patient required a new level of multitasking, all while suffering from profound sleep deprivation. My son had multiple undiagnosed food allergies, was continuously vomiting, and eventually developed failure to thrive. The guilt associated with not being home with him during this time was significant, as was the stress of taking him to multiple medical specialists, occupational therapy and nutritionists. Eventually, his health improved, and life calmed down. I went on to have two more healthy children, cathing throughout both pregnancies, delivering my middle son and then later my daughter again at 39 weeks - to the day! Aside from someone jokingly referring to me as "the perpetually pregnant one" I never felt judged in any way and was fortunate enough to find myself in a respectful and encouraging work environment.

Sara Trucco, MD

I had been interested in pursuing a career in pediatric interventional catheterization since medical school, but as a categorical fellow, I hesitated on committing to the career path because I was concerned that I would have to choose between being in the cath lab and starting a family. Since each pregnancy and any substantial maternity leave comprise around a year of time, I did not see an obvious route to be a successful interventional physician. All my interventional cardiology mentors were men, and while they were incredibly supportive of me, they could not advise me on these gender specific concerns. One of my mentors, Dr. Kanishka Ratnayaka, set up a phone call for me to talk to Dr. Sara Trucco who he mentioned had beautiful children while practicing as a pediatric interventional cardiology attending. I had read the available evidence on occupational radiation exposure in pregnancy but talking directly to someone with personal experience was invaluable in reassuring me that I would still have the choice to have a family while practicing.

My first pregnancy was towards the end of my interventional fellowship and first year as a new attending. I tracked my radiation exposure carefully and was selective about cases, when possible, with the knowledge that the total radiation dose and therefore occupational exposure for an infant patient is dramatically lower than for an adult. I was careful with shield placement in the laboratory, spent extra time collimating, and stepped back as far as possible during angiography (ALARA techniques that we should all be adhering to all the time). As Dr. Trucco had mentioned to me during that initial phone call, the challenges of postpartum return to work, pumping to facilitate breastfeeding and finding childcare for unpredictable hours have remained more stressful than those months of thinking carefully about fetal radiation exposure.

For many reasons, including the desire to spend more time taking care of my infant son, my second pregnancy occurred between faculty positions. The occupational related stressors were certainly less and the period before starting my next faculty position allowed for a length of maternity leave more akin to the rest of the world, albeit without pay. The stress of wondering whether I could still be successful in my chosen career however was significant. No one could reassure me that I would be accepted and effective in a procedural specialty after an extended period out of the cath lab as they had not seen examples of that path previously. I give credit to the interventional team at my current institution for being willing to consider that possibility and I hope it becomes a more accepted pathway for others in the future.

Elena Amin, MBChB

When I interviewed for my interventional fellowship in 1994, I was eight months pregnant with my first daughter. The fellowship was structured as a two-year fellowship. During the first year the "fellow" functioned as a clinical associate/junior attending helping with the inpatient service and did not go to the cath lab. The second year was entirely in the cath lab. That appealed to me since I was about to give birth.

Of course, I appeared very pregnant during the visit. After the first few minutes of my first interview, I was asked the following question: "Would you be interested in being a transplant doctor instead of an interventional cardiologist?"...The rest is history.

I can also speak to being pregnant four years later while on staff at the same institution as, guess what, an interventional cardiologist (I guess I was good enough to be hired). I was working as hard as always despite being pregnant, cathing, seeing patients in clinic, doing inpatient service, etc, etc, etc. At 34 weeks I developed a fever, 24 hours later I was in the hospital in labor, and 24 hours after that my blood culture came back positive for Listeria. Fortunately, my labor was stopped, and I was sent home with a PICC line on antibiotics (I told the doctors what to do with me after reading about 50 articles about Listeria in pregnancy and talking to the authorities in the field who wrote the articles). Two weeks later, at 36 weeks, my water broke, and I delivered a healthy baby girl.

Whether or not working as hard as I was working during my pregnancy had anything to do with my Listeria infection I will never know. I was actually reported to the CDC as one of about 20 cases of a Listeria outbreak in Ohio and Michigan related to Listeria contamination in a Sara Lee plant in Michigan. I do know that eating Sara Lee turkey breast sandwiches on most days, which took 30 seconds to prepare at home, was related to having no time to cook anything better, or to go to the

hospital cafeteria, as I was trying to get through my day and get home to see my four-year-old daughter.

I strongly believe that women in medicine who choose to have children should have better circumstances than we have in this country, perhaps during pregnancy, but even more importantly during the first few years of our children's lives, before they are old enough to attend school. We should look at some countries in Europe who do this better than we do. It would help the mental health of both mothers and children, and society as a whole. Something that as we all know is sorely needed at this moment in time.

I still have little time to do anything but medicine, but I'm not too far from retirement, and then could be talked into spending some time addressing some of these issues.
Lourdes Prieto, MD

I had my babies in 4th year interventional training and my first year as an attending. My male cath attendings would let me slip into the control room during angios, which was really nice since they didn't use any hanging lead shields! I tried to continue the practice of going into the control room when I was an attending and pregnant. Honestly, cases were much simpler and shorter back then. Nursing and pumping didn't seem to be an issue back then, but we weren't doing 6-hour transcatheter pulmonary valve replacement cases with pulmonary artery rehabilitation. I do remember eating a Wendy's triple cheeseburger, large fry, and frosty after one case, however, when I was pregnant with my son (who now doesn't eat red meat).

I got very good at balancing those breast pumps on the edge of my desk while I typed my notes. Now they have super cool pumps that you can use during a business meeting and no one knows! I shared an office with Luke Lamers, when I was a new attending, and he was an interventional fellow. He was so sweet to let me pump in there whenever I wanted. I would put a note on the door, and he would go hide somewhere else for a while. My husband would sterilize all of my equipment every night and pack it up for me for the next day in zip lock bags.

I only got six weeks of maternity leave, but that was good for me, especially with my first. I think I must have had post-partum depression and didn't know it. Going back to work was the only thing that brought me out of my funk, thinking I had ruined my life forever (my son was difficult in every way and is a joy now!). Going back to work showed me that the world was still going on, just as I had left it six weeks before (I thought maybe it had ended).
Aimee Armstrong, MD

From my perspective, working while pregnant was not a big issue, I felt great, enjoyed my pregnancies and always had the support from my male partners. But although I love my work, not having appropriate maternity leave was difficult especially having a toddler at home too. Coming back to work after six weeks with our physically and mentally demanding job seems like a challenge many of us have taken on without thinking much about it, but in my opinion it is wrong and there is a lot of room for improvement. In addition, having a private and always available room to pump would have been ideal! Something close to the cath lab that allows the opportunity to pump and maybe eat (not having to choose one or the other). We are the ultimate multitaskers, but this should not take a toll on our health (physical or mental) or on our amount of time shared with our families.
Jenny Zablah, MD

When I started my current job as a junior faculty straight out of fellowship, one of the senior cardiologist asked me directly if I was planning to get pregnant. I was shocked that he asked me directly when in reality you cannot ask that question. I had one child who was five years old but that just broke my heart. He said if you get pregnant then we won't have anyone in the cath lab.

It should be the individual person's decision as to whether to cath or not while pregnant. Nobody should dictate that.
Anonymous, MD

My son was born the first day of my chief residency and was one-year-old when I started cardiology fellowship. So, although I have not experienced cathing while pregnant, the decision to pursue a career in interventional cardiology has weighed heavily on the decision about when/if to have a second child. While my husband and I always envisioned two to three children, there never seemed to be a "right time" during cardiology fellowship to be pregnant. Once I decided to train in interventional cardiology, I was further deterred from getting pregnant out of fear of what people would think of me. I worried people wouldn't think I took my career seriously or that I wasn't dedicated. Now, having finished my training in my first attending job, I know there is never going to be a "right time." After months and months of trying to get pregnant, we are getting ready to start an IVF cycle. It is not lost on me that my older age is likely contributing to our difficulty in getting pregnant. Although I have extremely supportive colleagues, the perceived stigma around pregnancy and childbirth for women in intervention is an additional burden that women in this field have to shoulder relative to our male colleagues.
Anonymous, MD

As a woman who has gone through a few years of loss and fertility treatments, I experienced first-hand how stressful and heart-wrenching starting a family can be… not including the intersection with medicine and interventional cardiology. I reviewed the existing (limited) studies, and I completely understand when female interventionalists decide to cath throughout their pregnancies. However, given my own experiences, I decided to sit out of the cath lab when I found out I was pregnant, as the "unknowns" of radiation exposure were simply too much for me to accept. As a trainee, the guilt associated with this decision was compounded on the existing anxieties of desperately wanting a healthy pregnancy. Fortunately, I have supportive attendings who allowed me to delay my training for a year, giving me peace of mind to enjoy my pregnancy and the ability to be fully committed and guilt-free (perhaps just less guilty) next year as the interventional fellow.
Anonymous, MD

Being pregnant, as I always say, is part of our existence and cycle of life and we cannot be penalized for it. I have noticed that over the years there has been more acceptance, particularly from the younger generation. I have, however, seen both sides from the older generation. I was lucky to have Chuck Mullins as my mentor. I was pregnant three times during my training: once in residency, once in fellowship and once during my intervention year. The last one was a surprise. My fear of being discovered as pregnant led me to hide it for seven months. Double leading in silence and working long hours and going home to take care of my other two. When I finally could not hide it anymore, I was surprised to see the acceptance from Chuck. He said kindly and appreciatively, "Howaida you are the most productive fellow in every way," referring to my publications, of course. Frank Ing would say "Howaida you are a super doc, super mom and super woman!" All I can say is you can do it, and I am proud to be part of this team that is

fighting to make it easier for every woman that wants to pursue this beautiful career without fear or shame.
Howaida El-Said, MD, PhD

Concluding Remarks

Our field is small and with few interventional pediatric cardiologists at each institution, to allow true choice for the individual physician comes with significant challenges. Being out of the cath lab for the duration of pregnancy and providing a maternity leave beyond the minimum creates staffing challenges. We need creative solutions to these challenges for future generations so that we can promote success for women in our field, while maintaining true choice for each individual as it relates to their own health and their family.

With respect to trainees, there is voiced consensus amongst the women in pediatric interventional cardiology that the decision to be in the cath lab during pregnancy is an individual choice that will be respected without judgement. Many of us review the scientific data on occupational radiation exposure in pregnancy as part of the introduction to the cath lab at the start of categorical fellowship. This helps to allay anxiety if the issue subsequently arises for trainees. They are aware of who to discretely notify to receive fetal dosimeter badges if they choose to stay in the cath lab at a stage of pregnancy when they may not yet feel comfortable telling the wider team. They can also switch the timing of their rotations or feel confident about informing the team and feel supported in learning while participating only in the non-radiation-exposure components of the rotation. For those who find out they are pregnant after having already been exposed to radiation in the preceding weeks, being armed with scientific knowledge that the risks are, as far as we know, minimal, may provide some reassurance.

To promote gender equality and representation in our field we should continue to demonstrate that it is possible to work in the cath lab while pregnant if one chooses to do so. We also need to provide a staffing and training framework that allows interventional cardiologists to have a true choice regarding when and how to start their family.
Elena Amin, MBChB

References

1. Best PJ, Skelding KA, Mehran R, Chieffo A, Kunadian V, Madan M, Mikhail GW, Mauri F, Takahashi S, Honye J, Hernández-Antolín R, Weiner BH; Society for Cardiovascular Angiography & Interventions' Women in Innovations (WIN) group. SCAI consensus document on occupational radiation exposure to the pregnant cardiologist and technical personnel. Catheter Cardiovasc Interv. 2011 Feb 1;77(2):232-41. doi: 10.1002/ccd.22877. Epub 2011 Jan 28. PMID: 21061249.
2. Cheney AE, Vincent LL, McCabe JM, Kearney KE. Pregnancy in the Cardiac Catheterization Laboratory: A Safe and Feasible Endeavor. Circ Cardiovasc Interv. 2021 Apr;14(4):e009636. doi: 10.1161/CIRCINTERVENTIONS.120.009636. Epub 2021 Apr 20. PMID: 33877865.
3. Kneale GW, Stewart AM. Mantel-Haenszel Analysis of Oxford data. II. Independent effects of fetal irradiation subfactors. J Natl Cancer Inst. 1976 Nov;57(5):1009-14. doi: 10.1093/jnci/57.5.1009. PMID: 794502.
4. Dauer LT, Miller DL, Schueler B, Silberzweig J, Balter S, Bartal G, Chambers C, Collins JD, Damilakis J, Dixon RG, Marx MV, Stecker MS, Vañó E, Venkatesan AM, Nikolic B; Society of Interventional Radiology Safety and Health Committee; Cardiovascular and Interventional Radiological Society of Europe Standards of Practice Committee. Occupational radiation protection of pregnant or potentially pregnant workers in IR: a joint guideline of the Society of Interventional Radiology and the Cardiovascular and Interventional Radiological Society of Europe. J Vasc Interv Radiol. 2015 Feb;26(2):171-81. doi: 10.1016/j.jvir.2014.11.026. PMID: 25645407.
5. Durán A, Hian SK, Miller DL, Le Heron J, Padovani R, Vano E. Recommendations for occupational radiation protection in interventional cardiology. Catheter Cardiovasc Interv. 2013 Jul 1;82(1):29-42. doi: 10.1002/ccd.24694. Epub 2013 Mar 8. PMID: 23475846.
6. Quinn BP, Cevallos P, Armstrong A, Balzer D, El-Said H, Foerster S, Glatz AC, Goodman A, Goldstein B, Hainstock M, Janssen D, Kreutzer J, Latson L, Leahy R, Petit C, Shahanavaz S, Trucco S, Whiteside W, Zampi JD, Bergersen L. Longitudinal Improvements in Radiation Exposure in Cardiac Catheterization for Congenital Heart Disease: A Prospective Multicenter C3PO-QI Study. Circ Cardiovasc Interv. 2020 May;13(5):e008172. doi: 10.1161/CIRCINTERVENTIONS.119.008172. Epub 2020 May 15. PMID: 32408819.
7. Jain C. ACOG Committee Opinion No. 723: Guidelines for Diagnostic Imaging During Pregnancy and Lactation. Obstet Gynecol. 2019 Jan;133(1):186. doi: 10.1097/AOG.0000000000003049. PMID: 30575654.
8. Patel SJ, Reede DL, Katz DS, Subramaniam R, Amorosa JK. Imaging the pregnant patient for nonobstetric conditions: algorithms and radiation dose considerations. Radiographics. 2007 Nov-Dec;27(6):1705-22. doi: 10.1148/rg.276075002. PMID: 18025513.
9. McCollough CH, Schueler BA, Atwell TD, Braun NN, Regner DM, Brown DL, LeRoy AJ. Radiation exposure and pregnancy: when should we be concerned? Radiographics. 2007 Jul-Aug;27(4):909-17; discussion 917-8. doi: 10.1148/rg.274065149. PMID: 17620458.

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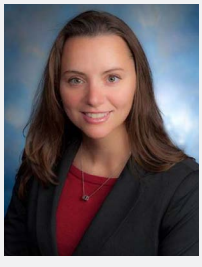

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CME Course on Lifelong Care of Adult CHD this Spring

Evidence-based updates on management advances and overcoming barriers to care

Patients with Adult Congenital Heart Disease (ACHD) are a rapidly growing and frequently underserved population. To equip clinicians to better meet their needs, Cleveland Clinic travels to Chicago in spring 2023 to offer a comprehensive 1.5-day CME course.

"Comprehensive Care for the Lifetime Treatment of Adult Congenital Heart Disease: A Case-Based Approach" will be held March 31st-April 1st at the InterContinental Chicago Magnificent Mile.

"Our goal is to update participants on clinical and research advances in the morphologies of congenital anomalies, the best diagnostic modalities and interventions, and the most appropriate management strategies for patients with ACHD," says course co-director Tara Karamlou, MD, MSc, a pediatric and adult congenital heart surgeon at Cleveland Clinic. "We aim to detail the highly complex ecosystem of congenital heart care in adults and describe how that care may be shaped by new approaches in management and recent guidelines."

30+ Faculty Over a Day and a Half

Over a full Friday and a Saturday morning, a faculty of more than 30 cardiologists, cardiothoracic surgeons and other clinicians with ACHD expertise from Cleveland Clinic and other top U.S. and international institutions will present practical, evidence-based updates on various aspects of ACHD care. After an opening session providing an overview of ACHD with patient perspectives, sessions will provide in-depth exploration of developments and innovations in the following:

- The pulmonary and tricuspid valves, including tetralogy of Fallot and Ebstein's anomaly
- Single-ventricle physiology
- The failing systemic right ventricle and systemic left ventricle
- Innovations in ACHD clinical care and research
- Special topics in the arc of care for ACHD

Each session includes presentations from multiple subspecialty perspectives, including imaging, diagnostic, surgical, interventional and electrophysiological.

"Recent advances have been made in various aspects of ACHD care, such as assessing and treating patients with pulmonary valve dysfunction and managing systemic right ventricular congenital disorders in adults, especially transposition of the great arteries," notes course co-director Hani Najm, MD, Chair of Pediatric and Congenital Heart Surgery at Cleveland Clinic.

"Moreover, as data mount on the management of adults who underwent Fontan surgery in infancy, the clinical care of these patients has improved," adds Joanna Ghobrial, MD, MS, Medical and Interventional Director of Cleveland Clinic's Adult Congenital Heart Disease Center. "However, not all clinicians are fully versed on these advances and insights, so we are focused on closing those knowledge gaps. We also will explore social and patient-perceived barriers that complicate the diagnosis and lifelong management of adult congenital heart disease."

New and Notable This Year

Notable in this year's course are presentations devoted to ACHD and socioeconomic disparities, ACHD and women's health, and ACHD and sports. Other highlights include an overview of groundbreaking research trials in ACHD, a patient panel with patient experience videos, and a keynote address by eminent congenital heart surgeon Vaughn Starnes, MD, who will also take part in a "fireside chat" with course co-director Lars Svensson, MD, PhD, Chair of Cleveland Clinic's Miller Family Heart, Vascular & Thoracic Institute.

"Lifelong surveillance is paramount for adults with congenital heart disease," says Dr. Svensson," and recent advances in imaging, surgical repair and catheter interventions have improved outcomes for these patients. We want to make sure awareness and utilization of those advances is as broad as possible."

Course and registration details are available at: ccfcme.org/achd23.

This activity has been approved for AMA PRA Category 1 Credit™.

This article is reprinted from Cleveland Clinic's Consult QD website, https://consultqd.clevelandclinic.org/course-on-lifelong-care-of-adult-chd-returns-to-chicago-this-spring/.

Transcatheter Aortic Valve Replacement Golden Years Are Over, Says GlobalData

Following Edwards Lifesciences CEO statement on the chilly outlook for transcatheter aortic valve replacement (TAVR)

James Spencer, Senior Data Scientist at GlobalData Healthcare, offers his view:

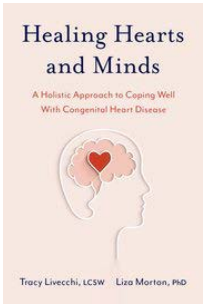
"For the past couple of years, TAVR has been enjoying an explosive growth. It was one of, if not, the fastest-growing cardiovascular markets, with consistent double-digit percentage growth of around 15%. However, with the aftereffects of the COVID-19 pandemic on the healthcare industry as well as the maturity of the TAVR market, the halcyon days of growth may be coming to an end.

"CEO Michael Mussallem reportedly said that 'the US TAVR procedure volumes continued to be impacted by regional staffing constraints, which were somewhat worse than we anticipated.' Edwards Lifesciences has now adjusted its growth forecast down to only mid-single digits, a far cry from the 10-18% in 2021. This is in line with GlobalData's own forecasts. The TAVR market has seen a growth of up to 40% in the last seven years as it exploded onto the scene with expanded indications and improving clinician knowledge of when and how to use the procedure. However, as the market matures, the scope for growth is ever diminishing.

"This is compounded by the macroeconomic factors at play in the US as well. The COVID-19 pandemic had a horrible effect on the healthcare industry. Short staffing and overwork due to a never-before-seen workload led to high levels of burnout and staff departure from the medical field. This in turn only exacerbated the issues, causing further staff to leave. Hospitals are struggling to find enough staff, and this is causing procedures to be delayed or even canceled. It will be a long time until the healthcare industry recovers from this blow, and until then, procedure volumes and, ultimately, patients will have to bear the consequences."

For more information, contact: pr@globaldata.com

The First Generation of CHD Survivors



Until 50 years ago, a diagnosis of **Congenital Heart Disease (CHD)** at birth or in childhood was a death sentence. Tracy Livecchi, LCSW, and Liza Morton, PhD, were among the first people in the world to receive **experimental, life-saving treatments** for their heart conditions as babies and children. Today, they are part of **the first generation of CHD patients that survived to adulthood.**

In ***Healing Hearts and Minds: A Holistic Approach to Coping Well with Congenital Heart Disease*** Livecchi and Morton share their personal stories of enduring traumatizing medical procedures and harrowing hospitalizations. While their survival – and the 75% increase in survival rates for CHD patients over the past few decades – are victories for modern medicine, both authors struggled to find mental health resources to help them overcome trauma and cope with the emotional toll of a lifelong medical condition. **"Healing Hearts and Minds" is the first psychological guide for CHD patients**, their families, and healthcare providers.

By sharing their personal stories and applying their experiences as mental health professionals, Livecchi and Morton seek to **close the gap in the lack of psychological resources for CHD patients** – both the first generation of survivors, and the next.

To order a copy, visit: https://global.oup.com/academic/product/healing-hearts-and-minds-9780197657287?q=healing%20hearts%20and%20minds&lang=en&cc=us

About the Authors

Tracy Livecchi, LCSW, is a Psychotherapist working in Connecticut and has been the Mental Health Consultant to the Adult Congenital Heart Association's Peer Mentorship Program for over 12 years.

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Healing Hearts and Minds: A Holistic Approach to Coping Well with Congenital Heart Disease

By Tracy Livecchi and Liza Morton will be published, in paperback, by Oxford University Press in January 2023 ($19.95 | 304 Pages | 9780197657287).

Powerful New Book Examines the Life and Work of Award-Winning Pediatrician

International Neonatologist, Researcher and Humanitarian Credited with Groundbreaking Medical Discoveries

By 2010, 4-7 million newborn babies each year required resuscitation at birth due to oxygen deficiency, and of those, one million had died. This is what has driven internationally renowned, award-winning pediatrician, neonatologist and researcher Ola Didrik Saugstad to fulfill his lifelong passion: to fight for newborn babies by questioning the prevailing practice of using pure oxygen to save the youngest and weakest among us.

In his new book, *Fighting for Air*, Saugstad reveals the struggles and triumphs, resistance and determination that led him to become one of the most sought-after experts in the field of neonatology. For 30 years, he fought to change the protocol for resuscitating newborn infants with ordinary air instead of pure oxygen, based on his research proving the latter can be a detriment to newborns. Despite rejection, stolen ideas and many uphill battles, his discovery that newborn babies should be given air instead of pure oxygen is now the guiding principle in hospitals throughout the world. The mortality rate among newborn babies suffering oxygen deficiency at birth has been reduced by 30% worldwide, with a range of 200,000-300,000 lives saved every year.

Set against the background of Saugstad's upbringing, family life and faith, *Fighting for Air* tracks in fascinating detail the factors that have influenced his firm resolve in improving the field of Neonatology. Outspoken against the "sorting society" and an advocate for the rights of myalgic encephalomyelitis (ME) patients, he shares uncommonly personal perspectives of how faith, politics and medical truths intersect, views that have informed his work from his early days as a medical researcher to becoming a celebrated pediatrician, professor and humanitarian.

The book examines the thoughts, beliefs and inspiration that informs every step of Saugstad's career. With a humble perspective and steadfast devotion to helping others, he has made it his life's mission to protect not only the youngest and most vulnerable, but to travel the world to save every life he can. Never faltering when faced with conflict, he credits his Christian faith as being crucial to his commitment to his work as well as being the beacon highlighting the power of reverence and the importance of family.

On what fuels his determination, Saugstad states: "What the outside world saw as personal ambition was something far more profound. It was a force over which I myself had no control, a power to push on and on. I had been given one of life's greatest and most challenging gifts: passion—zeal. Passion is a terrific force that changes lives—a gift and a responsibility handed to the few. It took many years before I realized that not everyone was like me. I had felt this passion ever since I was a little boy, whether as a soccer player, a high school pupil or as a medical student. I experienced the same when I attempted to come to the defense of those I viewed as debilitated. It was passion that saved me when I was under attack, and it was passion that kept me going whenever I encountered resistance."

Despite both breakthroughs and disappointments along the way, Saugstad's method of testing oxygen deficiency in newborn babies led to perhaps his biggest discovery. He found that the higher the presence of oxygen, the more oxygen radicals are formed, therefore causing greater damage. His research on hypoxanthine and free radicals has also contributed to the reduction of incubator blindness stemming from oxidative stress, a result of pure oxygen treatment in infants.

Fighting for Air inherently and effortlessly interprets the scientific into the comprehensible, reaching both the medical professional and the layperson. His profound knowledge of the world of medicine and the application of scientific research places him in an esteemed league like no other. With a steadfast moral backdrop, Saugstad also introduces the scientific processes used that led him in radical new directions few in his field pursue due to the sensitive nature of medical care for newborn infants. He is a firm believer that the uncritical use of oxygen for 200 years in the resuscitation of newborns is one of the greatest scandals in medicine, fueling many unnecessary tragedies.

About the Author

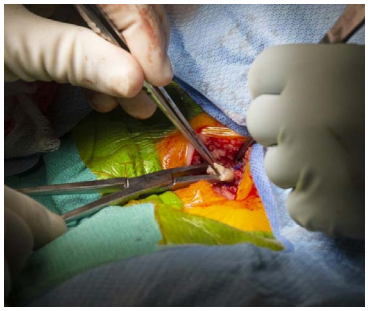
Ola Didrik Saugstad, MD, PhD, is a Norwegian pediatrician and neonatologist who is internationally recognized for his research on resuscitation of newborn children. From 1991-2018, he was Professor of Pediatrics at the University of Oslo and Director of the Department of Pediatric Research at the National Hospital. He has traveled all over the world giving lectures about newborn resuscitation and health and has been given numerous international and national awards, such as the Landmark Award by American Academy of Pediatrics (Perinatal section). Saugstad was nominated for the Nobel Prize in Medicine/Physiology for his research and is an appointed honorary member of a number of national associations and an honorary doctor and professor at international institutions. In addition to his engagement for the smallest among us, Saugstad has been involved in the debate about Biotechnology and Chronic Fatigue Syndrome.

Fighting for Air
Publisher: Waterside Productions
ISBN-10: 1958848034
ISBN-13: 978-1958848036
Available on Amazon

Baby Gets Heart Transplant with a Twist to Fight Rejection

Lauren Neergaard

Duke University doctors say a baby is thriving after a first-of-its-kind heart transplant -- one that came with a bonus technique to try to help prevent rejection of the new organ.



The thymus plays a critical role in building the immune system. Doctors have wondered if implanting some thymus tissue that matched a donated organ might help it survive without the recipient needing toxic anti-rejection medicines.

Easton Sinnamon of Asheboro, North Carolina, received his unique transplant last summer when he was six months old. But Duke waited to announce it until Monday after doctors learned the specially processed thymus implants appear to be functioning like they'd hoped -- producing immune cells that don't treat the tot's new heart like foreign tissue.

Doctors eventually will try weaning Easton off the immune-suppressing drugs required after a transplant, said Dr. Joseph Turek, Duke's chief of pediatric cardiac surgery.
The research is in very early stages and just one possible method scientists are testing in hopes of inducing what's called immune tolerance to a transplant.

But Turek says if it works, it could be attempted with other organ transplants, not just the heart.

Easton was a candidate for the experimental transplant because he had two separate health problems. He was born with some heart defects that surgeries right after birth failed to solve. And he suffered recurrent infections that doctors eventually realized meant his own thymus wasn't working properly.

Some babies are born without a thymus, which stimulates development of part of the immune system known as T cells. Separately, Duke researchers had been working with Enzyvant Therapeutics to develop lab-grown implants of donated thymus tissue to treat that rare disorder.

Easton got a combination of the two procedures. First surgeons implanted his new heart while the donated thymus was sent to a lab. About two weeks later, he had a second operation to implant the processed thymus tissue. His own partially working thymus was removed, to clear the way for new immune cells to take hold.

About six months later, testing shows the thymus tissue is building Easton well-functioning new T cells, said Turek.

MARCH

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PICS Society
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ISSN 1554-7787 print. ISSN 1554-0499 electronic.
Published monthly.