



PATTERNS OF PULMONARY CIRCULATION IN CHILDREN WITH PULMONARY ATRESIA - A STUDY OF TWO-HUNDRED CASES USING COMPUTED TOMOGRAPHY

Cardiology

Dr. Jomon Mathew John*	MBBS, MD, FIPC, Department of Paediatric Cardiology, Narayana Institute of Cardiac Sciences, Bangalore, India; Azeezia Medical College, Kollam, Kerala. *Corresponding Author
Dr. Viralam S. Kiran	MBBS DCH DNB FIPC, Senior Consultant Department of Paediatric Cardiology, Narayana Institute of Cardiac Sciences, Bangalore, India.
Dr. Venkatraman Bhat	MD, MNAMS, FRCR (Lon), Group Head Quality Radiology, Senior Consultant Radiology, Mazumdar Shaw Cancer Centre - Narayana Health, Bengaluru.

ABSTRACT

Objectives: To study the native pulmonary artery characteristics; the number, origin and course of the aorto-pulmonary collaterals and to characterise the associated intracardiac malformations in children with pulmonary atresia. **Methods:** A descriptive-retrospective study on children with pulmonary atresia who were evaluated in Narayana Institute of Cardiac Science (NICS), Bangalore India between 2011 and 2018 using computed tomography images and reports. **Results:** A total of 200 patients were studied with male/female ratio of 1.4. Pulmonary circulation provided predominantly by collateral arteries was seen in 33.5%, originating mainly from Descending Aorta with majority of collaterals supplying both lungs. Overall, 58% of patients with both Patent Arterial Duct and collaterals had these collaterals supplying the same lung perfused by the duct. The size of central Pulmonary Arteries had no correlation with the type of collateral vasculature. Short-segment pulmonary atresia was seen in 66.5% with confluent branch Pulmonary Arteries in majority of them. A total of 80 patients had complex anatomy, dominant of which was Double Outlet Right Ventricle (40%) and Corrected Transposition (25%), Coronaries were abnormal in 38 patients, commonest of which was origin from opposing sinuses (8/38). **Conclusion:** The pulmonary circulation in pulmonary atresia lacks predictability. A thorough evaluation of the pulmonary circulation is required in each case for the optimal strategy of management.

KEYWORDS

Pulmonary Atresia; Pulmonary Circulation; Major Aorto-pulmonary collaterals; Computed Tomography

INTRODUCTION

Pulmonary atresia is a malformation characterized by lack of luminal continuity, and absence of blood flow between right ventricle and pulmonary artery [1-3]. It either occurs with an intact interventricular septum or as a part of more complex cardiac malformations. In such malformations, an alternative route of blood supply to lungs becomes crucial for survival. The anatomical diversity that exists within this group of malformations deserves meticulous analysis and detailed assessment. A complete delineation of pulmonary vasculature along with the intracardiac malformation is essential for concrete planning of an optimal surgical strategy [4,5,6].

The pulmonary vasculature is divided into the pulmonary and bronchial systems. The intrapulmonary structures are supplied by the pulmonary arteries which regulate gas exchange as it branches into an extensive capillary network at the level of respiratory bronchioles and alveoli. Whereas the bronchial system perfuses the capillary bed within the bronchial wall and the structures of the perihilar region, including lymph nodes and the adventitia of elastic and large muscular pulmonary vessels [7].

A systemic blood supply to the lung was suggested by Galen in the Second Century AD. In 1732, Ruysch confirmed its presence and designated it as the bronchial artery. It is now known that the bronchial arteries may arise from the descending aorta, intercostal arteries, subclavian artery, or internal mammary arteries. The bronchial arteries may be classified as extra-pulmonary or intra-pulmonary. The extra-pulmonary artery gives off small branches to the oesophagus, mediastinal tissues, hilar lymph nodes, and the lobar bronchus. The intra-pulmonary bronchial artery distributes to the supporting tissue and structures of the intralobar bronchi, pulmonary pleura, lymph nodes, walls of the pulmonary artery, and to veins and nerves.

A close and important relationship exists between the pulmonary and bronchial vascular systems, which become most evident in the setting of aberrant cardiovascular development. Enlargement of the bronchial circulation is especially striking in association with congenital heart diseases associated with reduced pulmonary blood flow - such as pulmonary atresia or Tetralogy of Fallot. Furthermore, ligation of the pulmonary arteries produces enlargement and dilation of the bronchial artery resulting in the increase of pre-capillary anastomosis and the development of various abnormal flow routes [8].

Given the histological similarities and the temporal and spatial proximity of cardiac and pulmonary vascular development, it is likely that a common, multipotent mesoderm precursor exists that gives rise to both the heart and the pulmonary vasculature [9].

The available description of collaterals and pulmonary vasculature in pulmonary atresia is based on autopsy studies done on 31 specimens in 1985 and few angiographic studies on patients with Pulmonary Atresia with Intact Interventricular Septum (PA-IVS) or Pulmonary Atresia with Ventricular Septal Defect. The present-day imaging modalities offer superior clarity of the pulmonary circulatory patterns in patients with pulmonary atresia with underlying complex intra-cardiac malformations, which initial studies could not achieve. The present study was undertaken to delineate lung vasculature in children of pulmonary atresia with all the rare varieties included.

METHODS

Methodology

In the present retrospective study, data of 200 patients were collected from the medical records of children with Pulmonary atresia who were evaluated in NICS between the year 2011 to 2018. The Computed Tomography (CT) reports and images of these patients were analysed. Patient demographics such as the age, gender, weight and diagnosis were recorded and each data was further analysed based on the following characteristics

- Presence and Confluence of Pulmonary arteries
- The number, origin and course of the aorto-pulmonary collaterals
- Long segment vs short segment pulmonary atresia
- Associated Intra cardiac malformations
- Sidedness of the Aortic arch.

Statistical Analysis

Data were analysed with the software SPSS 22.0 version. Continuous variables were expressed as mean \pm SD and categorical variables as frequency or percentages. Value of $p < 0.05$ was considered as statistically significant.

Ethical Consideration

The study protocol was approved by the institutional ethical committee (NHH/AEC-CL-2018-324 dated 15th February 2019).

RESULTS

A total of 200 patients with pulmonary atresia were analysed. The frequency distribution of age and gender is depicted in table 1. Male to

Female Ratio was 1.4:1 (n=117/83). Overall, 72.5% (n=145) of study population was under-5-years age group.

Age groups	n (Total 200)	Female	Male	Percentage
1-30 days	6	0	6	3
1 month- 11 months 29 days	56	18	38	28
1- 5 yrs	83	39	44	41.5
5.1-10 yrs	35	16	19	17.5
10.1-15 yrs	11	6	5	5.5
>15 yrs	9	4	5	4.5
Total	200	83	117	100

When native pulmonary circulation is underdeveloped, vessels that represent primitive intersegmental arteries apparently continue into the intrapulmonary vasculature as Major Aortopulmonary Collateral Arteries (MAPCAs) which often originate from the descending Aorta, develop early in embryonic life and are tortuous with an unpredictable arborisation pattern. These intersegmental arteries normally regress with the establishment of central pulmonary to intraparenchymal artery connection. The MAPCAs are different from bronchial arteries and other native collaterals in numerous ways.

A collateral circulation is a naturally occurring artery-to-artery or arteriole-to-arteriole anastomoses that forms a network of specialised endogenous bypass vessels in healthy tissues. In vascular obstructive diseases, these collaterals serve to protect tissues from ischemic injury and exhibits minimal or no tortuosity (10,11), whereas MAPCAs do not form a nutritive plexus around bronchi. Unlike bronchial arteries, MAPCAs do not branch in their mediastinal course nor do they anastomose with intercostal arteries. They usually anastomose with the intrapulmonary arteries at or near the hilum.

A Patent Arterial Duct (PDA) becomes an important pulmonary blood supply in children with pulmonary atresia. It facilitates an adequate central pulmonary arterial size with a normal peripheral distribution. Thus MAPCAs are uncommon in that hemithorax perfused by the PDA and generally do not coexist (12)

The pulmonary vasculature in the study cohort had the patterns as depicted in table 2.

Pattern	Frequency	Percentage
1. No collaterals	1A. with PDA	72
	1B. without PDA*	
2.PDA with collaterals	2A. With MAPCAs	82
	2B. Only Tiny leash of collaterals	
3. Only MAPCAs	36	18
4.Only Tiny Leash of collaterals	10	5

* patients required surgical shunt

Among the 67 patients with MAPCAs, 80.6% had MAPCAs originating from Descending Aorta (n=54). The level of origin varied from T2 to T11, though T5 and T6 were frequently encountered, each representing 34 % of all MAPCAs (n=23 each). Seven had collaterals from Left subclavian Artery and another 7 from the Abdominal Aorta. Other sites of collaterals at a lesser frequency were: Arch of Aorta, Right subclavian Artery, brachiocephalic artery, bilateral subclavian arteries, distal arch of aorta, Left circumflex coronary artery and Right internal mammary artery.

Overall, 60% of these collaterals supplied both lungs (n=40), 27% supplied only the right lung (n=18) and 13 % supplied only left lung (n=9).

Among the 31 patients who had PDA with MAPCAs, 58% had major collaterals supplying the same lung perfused by the PDA (n=18).

It was also noted that both small diffuse and major collaterals co-existed in the same lung in 26 patients. The MAPCAs were present in all patients above 15 yrs (n=9) in comparison to patients less than 5 yrs with the same intracardiac malformation.

The intracardiac anatomy of the patients in this study cohort can be broadly classified into Simple and Complex anatomy. Patients with situs solitus, D-looped ventricles, levocardia, Pulmonary Atresia with

Intact Interventricular Septum (PA-IVS) or with a Ventricular Septal Defect (PA-VSD) were grouped under Simple Anatomy. Whereas additional defects to the above-mentioned lesion and abnormal situs were grouped under Complex Anatomy. Overall, 120 patients had simple anatomy with male to female ratio of 1.26:1 (n= 67/53), whereas complex anatomy was seen in 80 patients with a male preponderance (n= 50/30; M/F 1.66:1).

The most frequently encountered anatomy among the complex variant was Double Outlet Right Ventricle in 40% (n= 32/80). Table 3 depicts the frequency of each intracardiac anomaly associated with pulmonary atresia in our study.

Intracardiac Anatomy	Frequency	
Simple Anatomy	VSD	89
	Intact IVS	31
Complex Anatomy	Double outlet right ventricle	32
	Corrected Transposition	20
	AV Discordance	12
	Tricuspid Atresia	11
	DILV	9
	AVSD	6
	Ventriculo- Arterial Discordance	4
	Supra cardiac Total anomalous pulmonary venous connections	2
	Double inlet right ventricle	2
	Aortopulmonary window	2
	Atrioventricular criss cross connections	1
	Mitral atresia with hypo plastic left ventricle	1

* frequencies in complex anatomy are not mutually exclusive

Visceral situs was ambiguous in 8 patients and another 8 had situs inversus. Mesocardia was seen in 19 patients and 10 had dextrocardia. Coronaries were abnormal in 38 patients, of which 8 had coronaries arising from opposing sinuses. Conal branch of Right Coronary artery crossing Right Ventricular Outflow Tract (RVOT) was seen in 6 patients. Ectasia of coronary arteries was seen in 5 patients with simple intracardiac anatomy, of which 4 had intact PA-IVS and one with PA-VSD with this ectatic coronary giving few collaterals to both the lungs. Right Ventricle dependant coronary circulation was seen only in 3 patients with PA-IVS (n = 3/31).

Some of the other coronary anomalies observed at a lesser frequency are as follows: high origin of coronaries (n=3), Right Coronary Artery (RCA) crossing RVOT (n=3), Left Anterior Descending (LAD) from RCA (n= 2), common origin of coronaries (n=3), LAD coursing along Right Ventricular free wall (n=1), RCA from Left Circumflex artery (n=1) and from left coronary cusp via separate ostia (n=1), prominent RCA (n=1) and finally a coronary fistula to Main pulmonary artery (n=1).

Aortic arch sidedness refers to the corresponding bronchus crossed by the arch. In the present study cohort, left aortic Arch was seen in 169 patients whereas only 31 patients (15.5%) had right aortic arch (RAA). Sixty-one percent of patients with RAA had complex anatomy (n=19).

Majority (90.3%) of RAA had mirror image branching neck vessels, aberrant left Subclavian artery was seen in 2 patients and another one had a common origin of right innominate and left common carotid artery.

Patients with complex anatomy have a predominantly unifocal pulmonary vascular supply via PDA or tiny leash of collaterals along with it. The MAPCAs developed only in 23.7% of patients in this group (n=19), whereas 40 % of patients with simple anatomy developed MAPCAs (n=48).

The McGoon ratio is a practical method to estimate pulmonary arterial size (13) and is calculated by dividing the sum of the diameters of Right pulmonary artery at the level of crossing the lateral margin of vertebral column and Left pulmonary artery just proximal to its upper lobe branch by the diameter of aorta at the level of diaphragm.

McGoon ratio of each patient was compared with the variant of

collateral vasculature. The mean McGoon ratio in each subset was derived. [Table 4].

Table 4 : Mean McGoon Ratio in each vascular pattern

Variable name	category of vascular pattern	n	Male	Female	McGoon Ratio (Mean ± SD)	P Value
collateral pattern	Group 1A	51	31	20	1.7 ± 0.37	<0.001
	Group 1B	21	10	11	1.9 ± 0.29	
	Group 2	82	49	33	1.4 ± 0.39	
	Group 3	36	20	16	1.3 ± 0.41	
	Group 4	10	7	3	1.4 ± 0.33	

The ratio was found to be comparable among all the subgroups and the difference among the sub-groups was not statistically significant. In both genders, the least and the highest documented McGoon ratio was 0.6 and 2.5 respectively with a mean of 1.5.

Furthermore, the length of pulmonary atresia and the presence or absence of a confluence were meticulously looked for and documented. An atresia exists when there is a complete obstruction or lack of communication between ventricular mass and pulmonary arteries. It could either be due to an imperforate pulmonary valve or a muscular obstruction at the distal end of sub pulmonary infundibulum denoted as membranous and short segment atresia respectively [Figure 1].

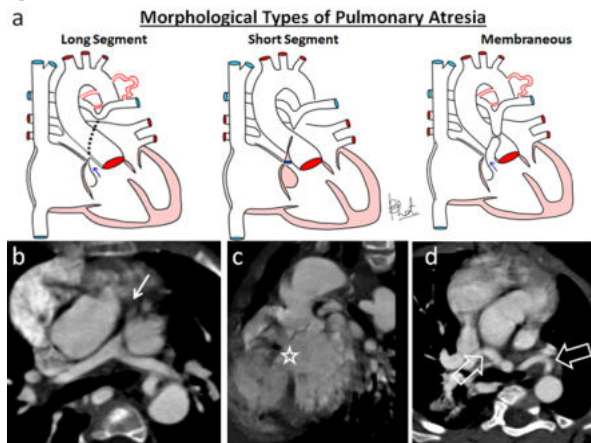


Fig 1: (a) Demonstrates the main types of pulmonary atresia. (b) Axial computed tomography Angiogram (CTA) demonstrates the atretic segment of main pulmonary artery (white arrow) (b) Long axis reconstruction shows blind right ventricular out flow and membranous VSD (star). (d) Axial view at a lower level demonstrates multiple large collateral arteries (MAPCA's) arising from descending aorta.

At times the muscular outlet septum fuses with the parietal musculature of right ventricle which makes the obstruction much more proximal and thus a long segment of atresia. In the present cohort, 66.5% had short segment of atresia (n=133) whereas membranous pulmonary atresia was seen in 14.5 % (n=29). Of the total 133 patients with short segment atresia , 72 had a simple anatomy. Long segment pulmonary atresia was seen in 19% (n=38) of the study cohort.

This trend of short segment being the most common followed by long segment and the least being membranous atresia was observed both in Simple and complex anatomy [Table 5] .Confluent branch pulmonary arteries were seen in all except 4 patients.

Table 5 : Frequency distribution of type of segment atresia

Segment of Atresia	Frequency	Simple	Complex
short segment	133	72	61
long segment	38	25	13
membranous	29	23	6
Total	200	120	80

DISCUSSION:

Pulmonary atresia with a VSD has a prevalence of 0.07 per 1000 live births (14,15). Pulmonary atresia with a complex anatomy represents a smaller fraction of an already rare condition. As a consequence, the existing literature has focused primarily on the former.

This retrospective study describes the variants of pulmonary vasculature in children with pulmonary atresia irrespective of their

underlying intracardiac malformations.

The complexity of pulmonary circulation determines the extent of surgical exploration necessary to perform unifocalisation, a surgical technique that eliminates extra cardiac sources of pulmonary blood flow and aims to incorporate as many pulmonary arterial segments as possible into a confluent pulmonary artery . A conduit is then placed from right ventricle to pulmonary artery confluence followed by VSD closure. The main determinants of postoperative right ventricular pressure which correlates with surgical outcome are adequacy of pulmonary vascular bed and pulmonary vascular resistance. For a satisfactory hemodynamic result, at least 10-16 lung segments need to be connected to the RV-PA conduit (16) Hence it is prudent that lung arborisation needs to be evaluated prior to surgery. If perfusion of a bronchopulmonary segment is contributed by both MAPCAs and native PAs, ligation of MAPCAs can be done. But if MAPCA is the sole supply, then that segment needs to be unifocalized.

Liao et al (3) studied 31 heart-lung autopsy specimens with pulmonary atresia where MAPCAs were seen in 20, amongst which 18 lacked a ductus and those who had one, didn't have major collaterals in 83% of the specimen (n=26). Hence an inverse relation between the two was drawn and postulated that even when ductus and MAPCAs coexisted in the same case, they supplied different lungs. In support of this view many other authors (17,18) published similar findings. However in our studied cohort, 58% of cases had MAPCAs and a ductus coexisting in the same lung.

Liao et al also documented the presence of a confluent central Pulmonary Arteries (PAs) in 87.5% of specimens with an identifiable pulmonary trunk whether atretic or patent. A similar if not stronger association was derived in our study with 98% having confluent branch PAs. Only one out of 4 patients had a short segment pulmonary atresia with non-confluent central PAs.

Liu et al (19) analysed 116 children with PA- VSD over a period of 1 year (2013-2014) to assess the efficacy of multi detector computed tomography (MDCT) over cardiac catheterization and transthoracic echocardiography in the evaluation of pulmonary vasculature. The authors reported MDCT to be most accurate test for delineation of MAPCAs and identification of native pulmonary arteries. They documented MAPCAs in 90.5 % of the studied population with majority supplying either right (41.4%) or left lung (45.1%).Confluent branch PAs were detected in 89.4% of patients and both branch PAs were absent in 34 patients. However, in the present study, MAPCAs predominantly perfused both lungs and confluent branch PAs was encountered in 98% of study cohort.

A definitive repair is possible in infants with good sized central PAs. Once central pulmonary arteries are formed during foetal life, the amount of blood flow through it is a key factor that determines its size and growth (18). Hence it is often necessary for patients with hypoplastic central pulmonary arteries to undergo an early palliative surgery to ensure progressive enlargement of arteries.

An average Mc Goon ratio is 2.1 in normal subjects. Ratio above 1.2 has an acceptable postoperative RV systolic pressure in Tetralogy of Fallot (20). A mean McGoon ratio of 1.5 was noted in our studied cohort and unlike many previously published articles and literature (18, 21) our analysis did not find a significant correlation between the size of central pulmonary arteries and the type of collateral vasculature.

A recent study conducted by Patric et al (22) on 50 patients with MAPCAs and atypical anatomy documented Unbalanced AVSD in 15 patients (30%) and another 5 (10%) had Complete AVSD. In our study AVSD was encountered only in 6 patients (3%) of which 4 (2%) had Unbalanced AVSD.

Overall, coronary anomalies accounted for 19% of study population, which is marginally higher than coronary anomalies found in a large single centre, retrospective analytic study of 2235 patients with Tetralogy of Fallot by Changela et al in 2010 (23). Authors have reported coronary anomalies in 15% of their cohort, and encountered 3 unusual coronary anomalies which includes a coronary cameral fistula to right ventricle, a giant coronary to PA fistula that arose from left anterior descending artery and an anomalous coronary from pulmonary artery.

Arch anomalies are often associated with Congenital heart disease (CHD). Based on the arrangement of arch vessels, the Edwards classification (24) describes 3 types: type I- RAA with aberrant left subclavian artery, type II -RAA with mirror image branching and type III RAA with isolation of left subclavian artery. Type I is the most common variation of right aortic arch (25) followed by type II which is strongly associated with CHD like Tetralogy of Fallot, Truncus Arteriosus, Tricuspid atresia and Transposition of great arteries with pulmonary stenosis (26). Whereas type I can be associated with normal intracardiac anatomy (27,28). In our study, type II prevailed as the majority.

Another European study (29) documents the outcome of 218 patients treated over a period of 26 years (1965-1991) in two leading cardiac centres in London, thereby modifying the disease course with state-of-the-art surgical management at that time. They were followed upto 40 years of age. Overall, 60 % of infants survived to 1 year, of which 65 % lived to 10 yrs. Only 16 % of these patients were alive at 35 years of age. Infective endocarditis (n = 17), stroke (n = 15) and RV failure (n = 16) were some of the cardiovascular complications encountered. Aortic regurgitation was recognised in 62% by the age of 30 years.

Survivors of pulmonary atresia after definite repair remain functionally well and less symptomatic but do face from time to time re-operations and therapeutic catheterisations for RV-PA conduit replacement and residual right ventricular outflow tract obstruction relief. A gradual deterioration of conduit function is observed due to loss of luminal diameter, peel formation and calcification. Loss of valve function in the conduit leads to pulmonary regurgitation, though well tolerated for years eventually leads to RV dysfunction and then LV dysfunction due to ventricle- ventricle interaction.

Patients unsuitable for a definitive repair are offered a palliative systemic to pulmonary artery shunt, develop progressive cyanosis and polycythemia as they survive into adulthood. A significant number of patients develop Aortic regurgitation (AR) as these shunts add to the Left Ventricular (LV) volume and therefore LV dilatation that results in aortic annular dilatation which worsens AR. A timely intervention with pulmonary valve or conduit replacement is expected to help reduce this risk.

CONCLUSION:

Pulmonary circulation influences the probability of repair and survival of patients with pulmonary atresia. Understanding the complexity and unpredictability of lung vasculature in these children, it is prudent that pulmonary vasculature must be evaluated thoroughly before offering the best possible management. Referral programmes needs to be reinforced to ensure early intervention and better outcomes.

REFERENCES

- Hofbecka M, Rauchb A, Leipold G, Singer H. Diagnosis and treatment of pulmonary atresia and ventricular septal defect. *Prog Pediatr Cardiol.* 1999; 9: 113–118.
- Tchervenkov CI, Roy N. Congenital heart surgery nomenclature and database project: Pulmonary atresia-ventricular septal defect. *Ann Thorac Surg.* 2000; 69:S97–S105.
- Liao PK, Edwards WD, Julsrud PR, Puga FJ, Danielson GK, Feldt RH. Pulmonary blood supply in patients with pulmonary atresia and ventricular septal defect. *J Am Coll Cardiol.* 1985; 6:1343–1350.
- Reddy VM, McElhinney DB, Amin Z, Moore P, Parry AJ, Teitel DF, et al. Early and intermediate outcomes after repair of pulmonary atresia with ventricular septal defect and major aorto-pulmonary collaterals: experience with 85 patients. *Circulation.* 2000; 101:1826–1832.
- Amark KM, Karamlou T, O'Carroll A, MacDonald C, Freedom RM, Yoo SJ, et al. Independent factors associated with mortality, reintervention, and achievement of complete repair in children with pulmonary atresia with ventricular septal defect. *J Am Coll Cardiol.* 2006; 47:1448–1456.
- Alwi M: Management algorithm in pulmonary atresia with intact ventricular septum. *Catheter Cardiovasc Interv.* 2006, 67: 679-686. 10.1002/ccd.20672.
- Shaw AM, Bunton DC, Brown T, Irvine J, MacDonald A. Regulation of sensitivity to 5-hydroxytryptamine in pulmonary supernumerary but not conventional arteries by a 5-HT(1D)-like receptor. *Eur. J. Pharmacol.* 2000; 408:69–82
- Liebow AA, Hales MR, Bloomer WE, Harrison W, Lindskog GE. Studies on the lung after ligation of the pulmonary artery. *Am. J. Pathol.* 1950. 26:177
- Millino C, Sarinella F, Tiveron C, Villa A, Sartore S, Ausoni S. Cardiac and smooth muscle cell contribution to the formation of the murine pulmonary veins. *Dev Dyn.* 2000; 218:414–25.
- Hillmeister P, Gatzke N, Dülsner A, et al.. Arteriogenesis is modulated by bradykinin receptor signaling. *Circ Res.* 2011; 109:524–533.
- Hecht N, He J, Kremenetskaia I, Nieminen M, Vajkoczy P, Woitzik J. Cerebral hemodynamic reserve and vascular remodeling in C57/BL6 mice are influenced by age. *Stroke.* 2012; 43:3052–3062.
- O'Leary PW, Edwards WD, Julsrud PR, Puga FJ. Pulmonary atresia and ventricular septal defect. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, eds. *Moss and Adam's heart disease in infants, children, and adolescents.* Baltimore, MD: Lip-pincott Williams & Wilkins, 2008:878–888.
- Nakata S, Imai Y, Takanashi Y, et al. A new method for the quantitative standardization of cross-sectional areas of the pulmonary arteries in congenital heart diseases with decreased pulmonary blood flow. *J Thorac Cardiovasc Surg* 1984; 88: 610-619
- Hoffman JI, Kaplan S. The incidence of congenital heart disease. *J Am Coll Cardiol.* 2002 Jun 19. 39(12):1890-900
- Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. *Circulation.* 2007 Jan 16. 115(2):163-72.
- Shimazaki Y, Tokusan Y, Lio M, Nakano S, et al. Pulmonary artery pressure and resistance late after repair of tetralogy of Fallot with pulmonary atresia. *J Thorac Cardiovasc Surg* 1990; 100:425-440
- Thiene G, Frescura C, Bortolotti U, Del Maschio A, Valente M. The systemic pulmonary circulation in pulmonary atresia with ventricular septal defect: concept of reciprocal development of the fourth and sixth aortic arches. *Am Heart J* 1981; 101:339-44.
- Bharati S, Paul MH, Idriess FS, Potkin RT, Lev M. The surgical anatomy of pulmonary atresia with ventricular septal defect: pseudotruncus. *J Thorac Cardiovasc Surg* 1975; 69:713-21
- J.Liu, H. Li, Z. Liu . Complete Preoperative Evaluation of Pulmonary Atresia with Ventricular Septal Defect with Multi-Detector Computed Tomography *PLoS One* 2016; 11(1): e0146380
- Piehlér JM, Danielson GK, McGoan DC, Wallace RB, Fulton RE, Mair DD. Management of pulmonary atresia with ventricular septal defect and hypoplastic pulmonary arteries by right ventricular outflow construction. *J Thorac Cardiovasc Surg* 1980; 80:552-567
- Jefferson K, Rees S, Somerville J. Systemic arterial supply to the lungs in pulmonary atresia and its relation to pulmonary artery development. *Br Heart J* 1972; 34:418-27.
- William L. Patrick, Richard D. Mainwaring, Olaf Reinhartz, et al. Major Aortopulmonary Collateral Arteries With Anatomy Other Than Pulmonary Atresia/Ventricular Septal Defect. *Ann Thorac Surg* 2017; 104:907–16
- V. Changela, S. Maheshwari, C. John. Unusual Cardiac Associations with Tetralogy of Fallot—A Descriptive Study. *Pediatr Cardiol* 2010; 31: 785–791
- Edwards JE. Anomalies of the derivatives of the aortic arch system. *Med Clin N Am* 1948; 32:925–48
- Türkvtan A, Büyükbayraktar FG, Ölçer T, Cumhur T. Congenital anomalies of the aortic arch: evaluation with the use of multidetector computed tomography. *Korean J Radiol* 2009; 10(2): 176–184.
- Glew D, Hartnell GG. The right aortic arch revisited. *Clin Radiol* 1991; 43(5):305–307.
- Berg C, Bender F, Soukup M, et al. Right aortic arch detected in fetal life. *Ultrasound Obstet Gynecol* 2006; 28(7):882–889
- Miranda JO, Callaghan N, Miller O, Simpson J, Sharland G. Right aortic arch diagnosed antenatally: associations and outcome in 98 fetuses. *Heart* 2014; 100(1):54–59
- Bull C, Somerville J; Ty E, Spiegelhalter D: Presentation and attrition in complex pulmonary atresia. *JAM COLL CARDIOL* 1995; 25:491-499.