Partial Anomalous Pulmonary Venous Return
Evaluation of 51 Cases

Filiz ŞENOCAK, M.D., Şencan ÖZME, M.D., Arman BİLGİÇ M.D.,
Süheyla ÖZKUTLU, M.D., Sema ÖZER, M.D.,
and Muhsin SARAÇLAR, M.D.

SUMMARY

Partial anomalous pulmonary venous return (PAPVR) is a congenital anomaly in which one or more, but not all, of the pulmonary veins are connected to a systemic vein or to the right atrium directly. Its incidence is higher in autopsy series than in clinical series.

We report 51 cases of PAPVR diagnosed by cardiac catheterization and evaluated from the aspects of age, sex, type and associated anomalies and diseases. (Jpn Heart J **35**: 43–50,1994)

Key words: Partial anomalous pulmonary venous return Associated anomalies

ABNORMALITIES of the pulmonary venous system are not common, although their real frequency is probably greater than that deduced from clinical and autopsy studies.[1] The first reported case of anomalous pulmonary venous connection was by Winslow in 1739. In 1942, Brody published a series of 106 cases.[2–5] In anatomic terms, the pulmonary venous anomalies may be classified as anomalous connections, stenotic connections and abnormal numbers of pulmonary veins.[1] Our subject is "Partial anomalous pulmonary venous return" defined as one or more, but not all, of the pulmonary veins being connected to the right atrium or to one or more of its tributaries. We studied 51 cases of PAPVR admitted to Hacettepe University Pediatric Cardiology Unit between 1972–1991 and diagnosed by cardiac catheterization. All patients were evaluated on the basis of age, sex, type of venous return and associated anomalies and diseases.

MATERIAL AND METHODS

Fifty-one patients with PAPVR diagnosed by cardiac catheterization at the Hacettepe University Pediatric Cardiology Unit between 1972 and 1991 were

From the Department of Pediatric Cardiology, Hacettepe University, Ankara, Turkey.
Address for correspondence: Filiz Şenocak, M.D., Kehribar Sokak 9/46–06700 Çankaya, Ankara, Turkey.
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evaluated. Cardiac catheterization and angiography were performed by the percutaneous or cut-down technique via the femoral vein. In two patients left heart catheterization was carried out via the femoral artery because of the presence of aortic coarctation. During catheterization, frequent oximetry samplings were obtained and shunt calculations were made subsequently. The diagnosis of PAPVR was established by direct injection of contrast material into the anomalous pulmonary vein or by examining the levophase of selective pulmonary artery injections. The diagnosis of PAPVR established by cardiac catheterization and angiography was confirmed during surgery in 33 patients.

Results

The findings from all patients and theraupetic interventions applied to them are summarized in Table I.

Of the 51 patients, 34 were male and 17 were female. The male/female ratio was 2/1. At the time of the diagnosis, their ages varied between 5.5 months and 26 years (mean: 9.11 years). Anomalous pulmonary veins originated from the right lung in 42 cases (82.3%), from the left lung in 5 cases (9.8%) and from both lungs in 4 cases (7.9%). The number of abnormally connecting pulmonary veins was one in 26 patients (50.9%), two in 18 patients (35.3%), and three in 7 patients (13.8%). There was a third pulmonary vein originating from the right lung in two cases (3.9%). In three cases there was an anomalous pulmonary venous confluence which drained in two cases into the right atrium and in one case into the innominate vein. The connection sites of anomalous pulmonary veins were as follows: Right atrium in 40 cases (77%), vena cava superior (SVC) in 7 cases (13.4%), innominate vein in 3 cases (5.8%) and persistent left superior vena cava in 2 cases (3.8%). In 47 cases (82.5%) anomalous pulmonary veins originated from the right lung; 39 (83%) drained into the right atrium, 7 (15%) into the SVC and one (2%) into the innominate vein. In 10 cases (17.5%) anomalous pulmonary veins originated from the left lung. Four (40%) drained into the right atrium, 3 (30%) into the innominate vein, 2 (20%) into the persistent left superior vena cava, and 1 (10%) into the SVC.

The PAPVR anomaly was isolated in only three cases (5.88%). The most commonly associated anomaly was atrial septal defect (ASD) which was seen in 41 (two ASD's in two cases) patients (80.39%). The second most common anomaly was persistent left superior vena cava which was seen in 6 cases (6.9%). The other associated anomalies and diseases in order of frequency were: In 5 cases (5.75%) ventricular septal defect (VSD), in 5 cases (5.75%) pulmonary stenosis (PS) (two infundibular, three valvular), and less frequently than these, tetralogy of Fallot (TF), double outlet right ventricle (DORV), congenital mitral

insufficiency, coarctation of the aorta (CA), dextrocardia, right arcus aorta, endocardial fibroelastosis (EFE), Ebstein anomaly, patency of the foramen ovale, subvalvular aortic stenosis, patent ductus arteriosus (PDA), single atrium, dilatation of the coronary arteries, stenosis at the pulmonary artery bifurcation, constrictive pericarditis, rudimentary SVC, pulmonary hypertension (40 mmHg systolic pulmonary artery pressure), renal agenesis and Hodgkin lymphoma. The types of atrial septal defects and their frequency were as follows: In 26 cases (60.47%) high venosum and in 17 cases (39.53%) secundum. In two patients there were two atrial septal defects, one of them high venosum, and the other of the secundum type. There was no primum type ASD in any case.

Nine patients failed to come to the control examinations after the catheterization. Thirty-seven patients were operated on (72.5%) and the diagnosis of PAPVR was proved in 33 of them. In the remaining 4 patients operated on, it was not possible to confirm the pulmonary venous connection according to the type of operation. In 30 patients with pulmonary-to-systemic flow ratios (Qp/Qs's) greater than 1.5 (81%) correction was made in order to direct the anomalous veins to the left atrium. One of these cases died shortly after the operation with signs of cerebral edema. The surgical techniques used in the other patients were devoted to correcting their most important cardiovascular anomaly.

The follow-up period ranged between 16 months to 20 years (mean: 5.24 years). Only one of the 41 cases followed (the 3rd case with TF) required another operation (left Blalock-Taussig shunt) two years later and transvenous pacemaker insertion for his atrial flutter 18 years after his second operation. The other patients remain asymptomatic with reasonable echocardiographic findings.

Discussion

The first case of PAPVR was reported by Winslow in 1739. The antemortem diagnosis of this anomaly using cardiac catheterization and angiocardiography was first reported by Dotter et al in 1949.[3,5]

PAPVR is the congenital anomaly in which one or more, but not all, of the pulmonary veins are connected to the right atrium or to a systemic vein, while the others drain into the left atrium. Obstruction of the pulmonary venous channels has not been described in this anomaly.[6] In spite of the fact that there is no difference of incidence between the two sexes, PAPVR was twice the incidence in boys in our series.[1]

In the developing embryo, the primitive foregut gives rise to the lungs, tracheo-bronchial tree and larynx. The primordial lung buds share a common vascular plexus (splanchnic plexus) with other derivatives of the foregut and, early on, drain through the common cardinal and umbilicovitelline veins. As develop-

Table I. The Results of Cases with PAPVR

Case No	Age (Years)	Sex	Number of abnormal veins R	L	Site of connection	Associated anomalies	Surgical procedure
1	8	M	1		Right atrium	Secundum ASD	Closure of the ASD
2	12	F	1		Right atrium	Secundum ASD	Discarded*
3	16	M		2	Coronary sinus (via persistent left SVC)	TF, persistent left SVC, right arcus aorta, ASD	Blalock Taussig shunt procedure (2 times)
4	14	F	2	1	Right atrium (separately)	High venosum ASD	Total correction
5	2	M		2	Right atrium	Situs inversus totalis, infundibular PS, VSD, ASD, persistent left SVC	Discarded
6	2.5	M	2	1	Right atrium (separately)	Secundum ASD	Total correction
7	10	M	1		Right atrium	Secundum ASD	Total correction
8	5.5/12 months	F	1		Right atrium	Secundum ASD, EFE	Total correction
9	8	F	1		Right atrium	Secundum ASD, Ebstein anomaly, right renal agenesis	Glenn procedure
10	9	F	2		SVC	High venosum ASD	Total correction
11	7	H	2	1	Innominate vein (via a common vein)	High venosum ASD	Total correction
12	6	F	1		Right atrium	Secundum ASD	Total correction
13	7	M	3		Right atrium (two of the pulmonary veins via a common vein)	Secundum ASD	Total correction
14	9/12 months	M	1		Right atrium	Dextrocardia, secundum ASD, DORV, VSD	Discarded
15	10/12 months	F	1		Right atrium	Secundum ASD, VSD, dextraposition of the aorta, right arcus aorta, PS	Discarded
16	4	M	1		Right atrium (separately)	DORV, secundum ASD, VSD, PS	Blalock-Taussig shunt procedure
17	2,5	M	1		SVC	Valvular PS	Pulmonary valvuloplasty
18	20	M	1		Right atrium	Secundum ASD, congenital MI	Total correction
19	26	M	2		Right atrium SVC	High venosum ASD, secundum ASD	Total correction
20	10	M	2		Right atrium (separately)	TF, secundum ASD	Total correction
21	10	F	2		Right atrium (separately)	High venosum ASD	Total correction
22	16	M	3		Right atrium (separately)	High venosum ASD	Total correction, died on the 1st day post operatively
23	6	M		1	Innominate vein	None	Exploration
24	13	M	1		SVC	Persistent left SVC, CA, PDA, subvalvular AS, dilatation of the coronary arteries	PDA ligation +aortic angioplasty

Case No	Age (Years)	Sex	Number of abnormal veins R	L	Site of connection	Associated anomalies	Surgical procedure
25	6	M	2		Right atrium (separately)	High venosum ASD	Total correction
26	6	M	1		SVC	DORV, PS, VSD, PA bifurcation stenosis	Discarded
27	9	M	2		Right atrium (separately)	High venosum ASD	Total correction
28	6	F	1		Right side of the common atrium	Common atrium, persistent left SVC	Total correction (with atrial septation)
29	12	M	1		Right atrium	High venosum ASD, cleft mitrale, mild PH	Discarded
30	13	F	2		Right atrium (separately)	Secundum ASD	Total correction
31	9	F	1		Right atrium	High venosum ASD	Discarded
32	6	M	2		Right atrium (via a common vein)	Rudimentary SVC, persistent left SVC, PFO	Total correction
33	8	F	1		Right atrium	TF (pink)	Total correction
34	4	M	2	1	Right atrium (via a common vein)	High venosum ASD	Total correction
35	4.5	F	2		Right atrium (separately)	Secundum ASD	Discarded
36	1	M	1		SVC	High venosum ASD	Total correction
37	5.5	M	1		Right atrium	High venosum ASD	Total correction
38	14	M	2		Right atrium (separately)	High venosum ASD	Total correction
39	10	F	2		Right atrium (separately)	Secundum ASD	Total correction
40	13	M	3		Right atrium (separetely)	High venosum ASD	Total correction
41	8	M	1		Right atrium	Secundum ASD	Closure of the ASD
42	14	F		1	Right atrium	Secundum ASD	Total correction
43	15	M	2		Right atrium (separately)	High venosum ASD	Discarded
44	14	M	1		SVC	None	Following clinically
45	7	M	2		Right atrium	Hodgkin lymphoma, constrictive pericarditis	Pericardiectomy
46	12	F	2		Right atrium (separately)	High venosum ASD, congenital MI	Total correction (including MVR)
47	10	M	2		Right atrium (separately)	Secundum ASD, high venosum ASD	Total correction
48	10	M		1	Innominate vein (via a vertical vein)	Secundum ASD, AC	Aortic angioplasty planned
49	8	M	1		Right atrium	Secundum ASD, persistent left SVC	Total correction
50	4	M	2		Right atrium (separately)	High venosum ASD	Total correction
51	8	M	1		Right atrium	None	Following clinically

* Discarded: Failed to return

ment proceeds, the splanchnic plexus differentiates into the primitive pulmonary vascular bed. At this stage of development (25–27 days' gestation) there is no direct communication with the heart. The right common cardinal system will later differentiate into the right horn of the sinus venosus, which will give rise to the right superior vena cava and azygous vein. The left common cardinal system will become the left horn of the sinus venosus and, with further differentiation, left superior vena cava and coronary sinus. The umbilicovitelline system will further differentiate into the inferior vena cava, ductus venosus and portal vein. At 27 to 29 days' gestation, the primitive pulmonary vein appears from the posterior superior wall of the primordial left atrium. At 28 to 30 days' gestation the pulmonary vein has engaged the pulmonary portion of the splanchnic plexus and begins to drain blood into the heart. The pulmonary portion of the splanchnic plexus begins to lose connections with the cardinal and umbilicovitelline systems at this time. With further development, tributaries to the pulmonary vein coalesce to form a common chamber, which is incorporated progressively into the posterior wall of the primitive left atrium. Complete incorporation results in two left and two right pulmonary veins entering through separate orifices. Variation in number of pulmonary veins is common. The most common variant (occurring in 24% of autopsy specimens) is the presence of a single pulmonary vein draining either the right or left lung. The second most frequent variation (occuring in 2% of specimens) consists of an extra pulmonary vein most commonly originating from the right lung.[5,6] In 3.9% of our cases there was a third pulmonary vein originating from the right lung. There were no fourth or fifth pulmonary veins, which were rarely encountered in our series.

Early atresia of the right or left portion of the common pulmonary vein while primitive pulmonary-systemic venous connections are still present, will lead to PAPVR.[10] Its incidence has been reported as 0.6–0.7% in autopsy series.[1,6] Blake et al observed 27 different anatomic variations of PAPVR.[7] In the literature, 70–90% of the cases show abnormally draining pulmonary veins originating from the right lung. In our series, we found 82.3% of abnormally connecting pulmonary veins to be of right lung origin, 9.8% of left lung origin and 7.9% from both lungs.

While left-sided pulmonary veins usually drain anomalously to derivatives of the left cardinal system, anomalous connections of the right pulmonary veins are mostly to derivatives of the right cardinal system. However, the embryologic splanchnic plexus is a midline structure, and this explains the development of possible crossed drainage of left-sided pulmonary veins to derivatives of the right cardinal system and vice versa.[1] There was no right sided pulmonary vein draining into derivatives of the left cardinal system in our cases. Meanwhile, of the left-sided anomalous pulmonary veins, 30% were connected to the innominate vein,

while 10% of anomalously draining left-sided pulmonary veins were connected to the SVC. Reviews of autopsy cases indicated the most common anomalous connections to be, in order of frequency: Right pulmonary veins to SVC, right pulmonary veins to right atrium and left pulmonary veins to left innominate vein.[1,5] Our observations of drainage localization of anomalous pulmonary veins showed that 83% of right sided pulmonary veins were draining into the right atrium, 15% into the SVC and 2% into the innominate vein. Of the left sided pulmonary veins, 40% were draining into the right atrium. It is an interesting result, because in the literature, there were almost no reported cases of anomalous left-sided veins draining into the right atrium.[8] Thirty percent of the left-sided anomalous pulmonary veins were draining into the innominate vein, 20% to the persistent left superior vena cava and 10% to the SVC. There was no abnormally draining pulmonary vein to the inferior vena cava (IVC) in the series.

Usually, PAPVR is found in association with other cardiac defects, most commonly an ASD of the secundum or sinus venosus type. Primum ASD is encountered rarely in this type of anomaly. Similarly, PAPVR occurs in about 10% of patients with ASD.[1,5,8] In our series, 80.39% of the associated cardiovascular anomalies were atrial septal defects. Of the atrial septal defects, 60.47% were high venosum type and 39.53% were secundum type. There was no primum type ASD.

PAPVR with intact interatrial septum, which has been rarely reported in the literature, occurred in 19.6% of our cases.[7,9,10] Of all the patients, 5.88% had isolated PAPVR anomaly.

Most patients with PAPVR with or without an interatrial septum defect do not exhibit symptoms in early life. Patients are often referred with a cardiac murmur or an abnormal chest roentgenogram. The most common complaint is mild exercise intolerance. Progressive symptoms usually begin in the mid-thirties or early forties. The findings are usually typical of an uncomplicated ASD. Respiratory symptoms and recurrent pneumonia may be observed in patients with scimitar syndrome in which the right-sided pulmonary veins drain into the IVC because of pulmonary parenchymal anomalies. In our series, there was no patient with scimitar syndrome. Patients with PAPVR and associated cardiac defects usually have signs and symptoms related to the associated defect.[1,5] Associated anomalies reported with PAPVR are: Dextrocardia, azygous continuation of the IVC, congenital mitral stenosis or atresia, DORV, VSD, FT, PS, CA, PDA, aortic stenosis and hypoplasia of the aorta.[5] Associated anomalies and diseases of our cases are listed in Table I. The second most common anomaly after ASD was persistent left SVC, followed by VSD and PS.

The radical therapy in PAPVR anomaly is surgery. In the presence of a hemodynamically significant left-to-right shunt (QP:QS greater than 1.5–2) one

should choose surgical therapy. Associated major cardiac lesions may alter this recommendation. Surgical mortality in patients without pulmonary hypertension is below 1%.[1,5,6,8] Only one of our patients had pulmonary hypertension, but this case left before surgery. The 37 patients operated on made up 72.5% of all the cases. The surgical approach was carried out in order to direct the anomalous pulmonary veins into the left atrium in 81% of the operated patients. In the other cases surgical therapy was aimed at correcting their major cardiovascular anomaly. One of the operated cases (2.7%) died shortly after surgery with signs of cerebral edema. Other cases followed up are in good condition.

In conclusion, we emphasize the importance of ruling out PAPVR in patients with congenital heart defects in order to avoid surgical complications.

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