Severe Pulmonic Stenosis with Intact Ventricular Septum and Right Aortic Arch

By Walter J. Gamble, M.D., and Alexander S. Nadas, M.D.

It has been assumed that patients with pulmonic stenosis and a right aortic arch practically always have a ventricular septal defect as well. As a matter of fact, in the differential diagnosis of pulmonic stenosis and right-to-left shunt, the presence of a right aortic arch often was taken as conclusive evidence of a tetralogy of Fallot-type lesion.1-3

A review of the literature revealed among several hundred, one patient with the presumptive diagnosis of pure pulmonic stenosis, who had a right aortic arch.4,5 This diagnosis was not made preoperatively, and the evidence is exclusively a surgical one, based on the findings at closed valvulotomy, and on a favorable postoperative course. Taussig reported having seen one other patient with valvular pulmonary stenosis, intact ventricular septum, and a right aortic arch, but did not give details.6

The three cases to be reported have been studied and operated upon at the Children's Hospital Medical Center between January 1960 and June 1963. Conclusive evidence is presented that all three had severe pulmonic stenosis, a right aortic arch, and intact ventricular septum.

Case Reports

Case 1

A heart murmur was discovered in this boy (R.M.) at 2 months of age, when he had pneumonia. At 24 months of age, when first seen at the Children's Hospital Medical Center, he had a grade V, long systolic, ejection murmur. His electrocardiogram showed an axis of +120° with right ventricular hypertrophy. Cardiac fluoroscopy showed the heart to be of normal size, but the configuration was consistent with concentric right ventricular hypertrophy. The aortic arch was noted to be on the right, and an oblique filling defect in the esophagus was interpreted as representing an anomalous left subclavian artery passing from the right, behind the esophagus upwards to the left (fig. 1A).

By 6 years of age, a slight decrease in exercise tolerance was reported, and slight cyanosis and clubbing were noted.

At 11 years of age, there were definite cyanosis and clubbing. A precordial bulge was noted. The second heart sound was single and moderately decreased in intensity. A grade III, long, ejection murmur was heard loudest at the third left interspace, and was transmitted to the entire precordium. The electrocardiogram showed an axis of +115°, "P pulmonale," and severe right ventricular hypertrophy (fig. 2, top). A chest x-ray showed possible slight cardiac enlargement (fig. 3A). The main pulmonary artery region was concave.

Cardiac catheterization was carried out at 11 2/12 years (table 1) and showed infundibular pulmonic stenosis. Right ventricular pressure exceeded systemic pressure. A right-to-left shunt was demonstrated at the atrial level. No shunt at the ventricular level could be demonstrated by means of oxygen saturation data, cineangiograms, nor by dye-dilution curves, with injection of indo- cyanine green into the right ventricle and sampling from the brachial artery.

At 11 6/12 years, during total cardiopulmonary bypass, a patent foramen ovale was sutured. Through a ventriculotomy, the infundibular stenosis, composed of fibromuscular tissue and diaphragmatic in nature with two small holes 3 and 4 mm. in diameter, respectively, was resected. The pulmonary valve was not stenotic. No ventricular septal defect was noted. The patient required digitalis postoperatively, and made an un-
eventful recovery. He has been asymptomatic since surgery.

At 14 4/12, (2% years after operation) he was restudied by cardiac catheterization. Auscultation revealed no significant murmurs. The heart sounds were of normal intensity. The second heart sound demonstrated a normal variation in splitting with respiration. Right ventricular systolic pressure was 22 mm. Hg. No significant pressure gradient was found across the right ventricular outflow and pulmonary valve regions. No shunting of blood could be detected by oxygen saturation data, cineangiography, or hydrogen appearance times.

Case 2

A murmur and enlarged heart were discovered on this patient (P.R.) at 10 weeks of age, when she was hospitalized for croup and pneumonia. A grade III to IV systolic murmur was heard when she was first seen at the Children’s Hospital Medical Center at 2 2/12 years. No evidence of cyanosis was noted. An electrocardiogram showed an axis of +70°, rSR' pattern in the right precordial leads, and voltages within normal limits. Cardiac fluoroscopy showed considerable enlargement, with right ventricular configuration. An aberrant left subclavian artery was noted (fig. 1B).

By 6 9/12 years, slight decrease in exercise tolerance was reported. At 9 3/12 years “P pulmonale” and definite right ventricular hypertrophy appeared.

At 10 9/12 years she was found to have a single second heart sound and a grade IV, long, systolic murmur at the second left interspace. This was thought to be pansystolic at the lower left sternal border. Cardiac catheterization (table 1) demonstrated severe infundibular pulmonic stenosis and a right aortic arch. No evidence of a ventricular septal defect could be found by oxygen saturation data nor by cineangiography with multiple injections into the left and right ventricles. Right ventricular systolic pressure ranged from 175 to 225 mm. Hg systolic, well above systemic arterial pressure. There was a 45 to 50 mm. rise in pressure following premature beats. Peripheral arterial blood was fully saturated.

At 11 8/12 years of age, with slightly increasing exercise intolerance, she was subjected to surgery. Slight, but definite tricuspid regurgitation was noted. During total cardiopulmonary bypass with cold arrest, marked infundibular obstruction consisting of fibromuscular tissue was found through a ventriculotomy and was excised. No valvular stenosis was noted. A careful inspection of the ventricular septum, especially from the crista region well down to the tricuspid valve, revealed no septal defect. The aorta was noted to be on the right. Her postoperative recovery was uneventful. Six months later there was partial regression of the right ventricular hypertrophy by electrocardiogram, with disappearance, of the “P pulmonale.” A chest x-ray showed reduction in the heart size. Twelve months postoperatively the second heart sound was noted to show normal

Figure 1

Vascular anomaly as demonstrated by barium swallow, showing filling defect caused by aberrant left subclavian artery posterior to esophagus. A. Case 1. B. Case 2.
Case 3

Shortly after birth this infant (R.N.) became ashen and cyanotic. No murmur was found, and chest x-ray was reported as normal. He was transferred to the Children's Hospital Medical Center at 10 days of age. He was noted to be acyanotic at rest, but became dusky and blue when feeding. A tapping right ventricular impulse was felt. The second heart sound was thought to be single. An inconstant click was heard at the lower left sternal border. A grade II to III ejection murmur was heard at the second and third intercostal spaces and transmitted down to the lower left sternal border. An electrocardiogram, at 11 days of age, showed an axis of +135° and right ventricular hypertrophy, probably abnormal for age. Cardiac fluoroscopy at 4 weeks showed moderate cardiac enlargement, with right ventricular configuration (fig. 2C). The aortic arch was noted to be on the right, and the pulmonary vasculature was slightly decreased. Ear oximetry showed 79 per cent saturation at rest, 70 per cent on crying. At 6½ months rather sudden deterioration, including increased cyanosis, difficulty in feeding, and decreased exercise tolerance, was noted. Examination disclosed marked cardiac enlargement, with a right ventricular impulse. The second sound was felt to be single. A grade III ejection murmur was heard. The liver was down 3 cm. An electrocardiogram showed a mean frontal plane axis of +135°, increased right ventricular hypertrophy, and "T pulmonale" (fig. 2, bottom).

Cardiac catheterization (table 1) was carried out at 7 months. This suggested severe infundibular and valvular pulmonic stenosis with right ventricular systolic pressure well above systemic level, ranging from 140 to 160 mm. Hg. One and a quarter hours after intravenous digoxin the right ventricular systolic pressure was 210 mm. Hg. A right-to-left shunt was demonstrated at the atrial level by oxygen saturation data, ascorbic acid appearance times, and by selective cineangiography. No shunt could be demonstrated by the latter two methods at the ventricular level. Cineangiograms also suggested peripheral pulmonic stenosis at the junction of the right and main pulmonary arteries.

Three days later the patient underwent transventricular valvulotomy. The main pulmonary artery was too small (4 to 5 mm. diameter) to permit a transarterial approach to the valve. The

Table 1

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( )—mean pressure, *not entered.
postoperative course was uneventful. He was continued on digitalis. Two months postoperatively he was much more active, was feeding well, and was virtually acyanotic. A grade III to IV long, systolic, ejection murmur was present, maximum at the second left intercostal space, transmitted to the neck, lower left sternal border, and back. There was a suggestion of a faint, early protodiastolic blow. The second heart sound continued to be single, and the heavy right ventricular impulse was still present. The electrocardiogram showed some decrease in right ven-

Figure 2

Electrocardiograms before and after operation.

Figure 3

Preoperative x-rays of patients. A. Case 1, age 11 7/12 years. B. Case 2, 10 6/12 years. C. Case 3, age 7/12 years.
tricular potentials, and "P pulmonale" persisted. The chest x-ray showed the heart to be slightly larger than preoperatively. Oxygen saturation, by ear oximetry, had increased from 58 per cent before operation to 86 per cent.

Discussion

There is nothing unusual in the family and prenatal history of the first and second patients. The gestation of the third patient was complicated by bleeding during the second month, and the mother was treated with oral norethindrone, 5 mg. twice a day for 15 days.

There is a striking similarity between cases 1 and 2. The same vascular anomaly was noted in both (fig. 1), suggesting an identical embryologic mechanism in these two patients. The stenosis was proved to be infundibular, and was described as diaphragmatic at surgery in case 1, and appeared to be the same by angiography in case 2. Because of the "blind" surgical approach, and less than completely satisfactory angiograms, no definite localization of the stenosis is possible in case 3. The surgeon had the impression that the obstruction was at the valve level.

After surgery all three patients showed striking clinical improvement. A virtual disappearance of the pressure gradient across the right ventricular outflow tract was demonstrated at catheterization in patient 1 (table 1), a satisfactory drop in pressure in patient 2 was implied in the postoperative electrocardiographic changes. Not enough time has elapsed to assess patient 3 in this respect.

The clinical picture, particularly the appreciable cardiomegaly (fig. 3) and the electrocardiogram (fig. 2), as well as the right ventricular pressure well above systemic arterial level, suggested preoperatively the presence of pulmonic stenosis with an intact ventricular septum. On the other hand, the diagnosis of pulmonary stenosis with ventricular septal defect was implied by the finding of a right aortic arch. The murmurs were all described as long ejection murmurs, maximal at the second (and third) left intercostal space at the left sternal border. Cases 1 and 2, in addition, also had pansystolic murmurs at the lower left sternal border which has been thought to reflect tricuspid regurgitation.

At catheterization the behavior of the right ventricular pressure pulse (absence of notch on upstroke) particularly after premature contractions was in favor of an intact ventricular septum, while the deep peripheral cyanosis and the finding of infundibular stenosis favored the presence of a ventricular septal defect. Indicator-dilution curves and cineangiograms were the principal means by which the presence of a ventricular septal defect was excluded preoperatively. Surgical observations and postoperative course substantiated the clinical assessment.

The conclusion to be drawn from these three cases is simply that infundibular and possibly valvular pulmonic stenosis with an intact ventricular septum and a right aortic arch does indeed exist, although obviously it is very rare (less than 1 per cent of patients with pure pulmonic stenosis). If clinical and physiologic evidences point to the diagnosis of pulmonic stenosis with an intact ventricular septum, the presence of a right aortic arch should not negate this conclusion. Obviously more than average care should be taken in these instances to exclude the presence of a ventricular septal defect by means of indicator-dilution curves and selective angiograms.

Summary

Three cases with pulmonary stenosis, intact ventricular septum, and right aortic arch are presented, along with physiologic data. Two cases had infundibular stenosis and, in addition, an aberrant left subclavian artery. The third case was thought to have valvular stenosis. All three patients have been successfully operated upon and show good clinical results.

Acknowledgment

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References


**Stephen Hales 1677-1761**

Stephen Hales, the clergyman of Teddington (1677-1761), who was particularly interested in plant physiology, made significant observations on the exchange between air and blood. Lavoisier later said that he was “the first who examined the problem from the quantitative point of view; he developed several devices, simple and easy to handle, in order to measure exactly the volume of air.” In Vol. I of his *Statistical Essays* (1731), Hales described the apparatus making it possible to “take an estimate of the quantity of Air absorbed or fixed or generated by the breath of living animals.” In Experiment CVII he measured the amount of air absorbed by breathing, and in Experiment CX he defined the site of absorption, marveling at the vast expanse of the alveolo-capillary membrane:

“. . . but some of the elasticity of air which is inspired is destroyed and that chiefly among the vesicles . . . whence probably . . . acid spirits . . . are conveyed in the blood which we see by an admirable contrivance spread into a vast expanse commensurate to a large surface of air from which it is parted by very thin partitions; so very thin as thereby probably to admit the blood and air particles . . . within the reach of each other’s attraction, whereby a continued succession of fresh air must be absorbed by the blood.”—André Cournand, M.D. *Circulation of the Blood*. Edited by Alfred P. Fishman, M.D., and Dickinson W. Richards, M.D., New York, Oxford University Press, 1964, p. 38.
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