VENTRICULAR SEPTAL DEFECT WITH NATURALLY ACQUIRED INFUNDIBULAR OBSTRUCTION—A REPORT OF THREE CASES

By T. F. Loh, M.D., M.B., B.S., M.R.C.P. Ed., D.C.H.
(Department of Paediatrics, University of Singapore)

In a series of 298 cases of ventricular septal defects, it is noted that one of the ways in which the patient improves spontaneously, is by the natural acquisition of infundibular obstruction.[4] Hypertrophy of the right ventricular outflow tract occurs under the prolonged stress of high pulmonary blood flow and pulmonary artery pressure, giving rise to a distinct pressure gradient between the right ventricular chamber and the pulmonary artery. When the gradient is sufficiently great, it causes a relative stenosis which progresses with time. When it becomes severe, the obstruction becomes effective in reducing the left-to-right shunt across the ventricular septal defect by raising right ventricular pressure, thus protecting the pulmonary circulation. As a result, the patient improves and becomes practically asymptomatic.

The incidence of the natural acquisition of infundibular obstruction is not known but it is generally observed to be not uncommon.

If the hypertrophy increases even more, the obstruction becomes so severe, that the pressure in the right ventricle equals that of the left, and there is practically no flow across the ventricular septal defect. At this point when both ventricular pressures are balanced equally, any increase on the right side will cause a reversal of the shunt giving rise to cyanosis. This commonly happens after any form of physical exertion. Cyanosis may even persist at rest, if the degree of obstruction is severe enough. Thus, a situation like that of the Fallot's tetralogy is created.

In the present series of 298 patients investigated, three of them have acquired severe infundibular obstruction confirmed by serial cardiac catheterization. Their case histories are as follows:—

Case 1. N.P.Y. 5 years female. No. P. 60143

She was the youngest of three siblings. Her birth history was normal and birth weight was 7 lb. She was first admitted at the age of 4 months as an emergency case of chest infection and cardiac failure. These symptoms were recurrent and she was hospitalised seven times during infancy, and a total of ten times during the first eighteen months of her life. Cyanosis was never noted during this period. Clinically, she had ventricular septal defect with pulmonary hypertension which was confirmed by cardiac catheterization at seventeen months.

Her case was discussed at the weekly Cardiac Conference. In view of her history of recurrent chest infections and intractable cardiac failure, banding of the pulmonary artery was strongly recommended as a palliarive measure by Professor Roy, Visiting Professor of Cardiology in the University of Singapore. However, this procedure was not carried out chiefly because of the inexperience at that time of the cardiac surgeon on this technique.

She was treated conservatively, and surprisingly, began to show improvement. Episodes of chest infection and cardiac failure became less frequent and less severe. She had two further admissions for bronchitis at $2\frac{1}{2}$ years and 4 years of age.

She was followed-up closely in the Cardiac clinic and was thriving satisfactorily from 2 years until $4\frac{1}{2}$ years of age, when her mother noticed blueness around the lips and in the extremities after exertion. There was no history of squatting. Her effort tolerance was fair but she had to rest after moderate exertion.

On examination, she was well-nourished, weighing $29\frac{1}{2}$ lbs. She had cyanosis with slight clubbing of the fingers. Her pulse was 90/min., regular and of good volume, BP = 100/80. Her heart was not enlarged. There was a precordial bulge with a systolic thrill and an ejection systolic murmur at the 3rd left intercostal space at the sternal edge. There was a single pulmonary second sound, probably due to the aortic valve closure, Hb. = 14.2 gm.%, PCV = 45%.

Her electrocardiographic changes are as shown in Fig. 1.

The electrocardiogram done at $1\frac{1}{2}$ years showed a mean QRS axis of +45° with an anticlockwise loop, left ventricular diastolic overload in V_6 and right ventricular hypertrophy as suggested by the upright T wave in V_1. The electrocardiogram done $3\frac{1}{2}$ years later showed a change of the mean QRS axis to +135° and isolated right ventricular hypertrophy.

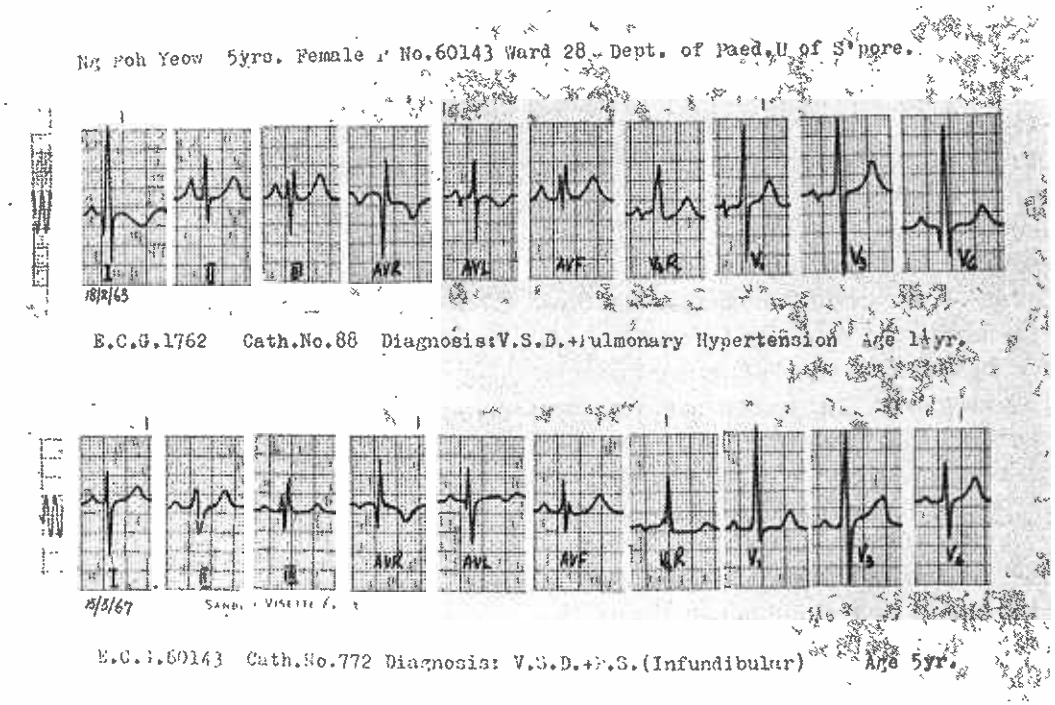


Fig. 1. Electrocardiograms done at 1½ years of age show the change from a biventricular hypertrophy to an isolated right ventricular hypertrophy.

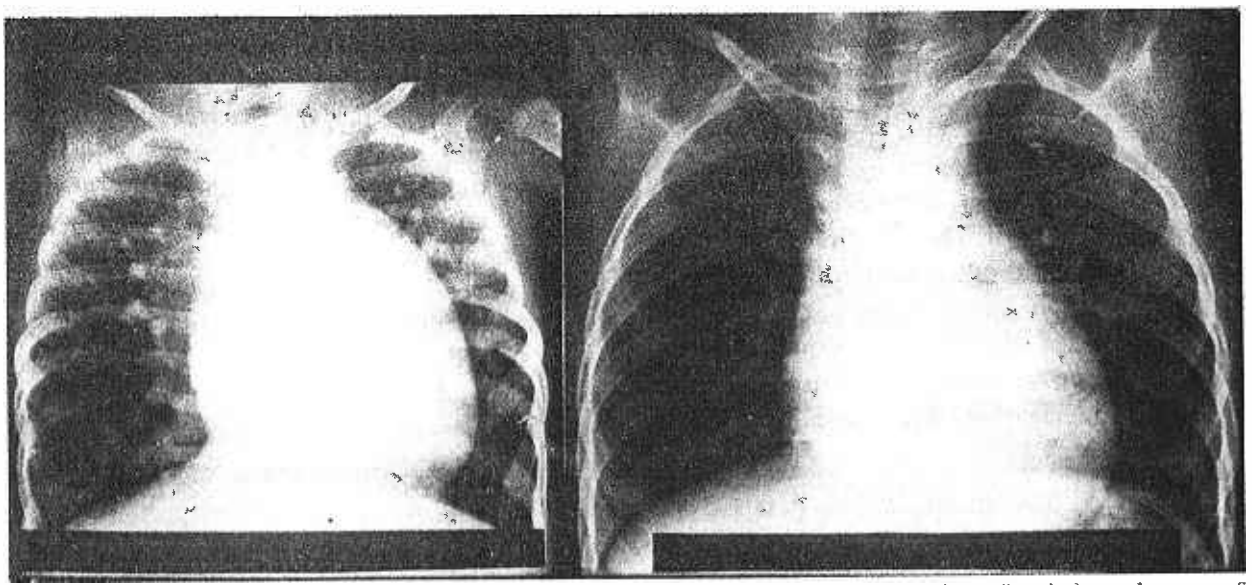


Fig. 2. The chest X-ray on the left shows marked pulmonary plethora (L-R shunt of 6.4 L/min.) at the age of 1½ years and that on the right shows oligaemic lung fields at 5 years.

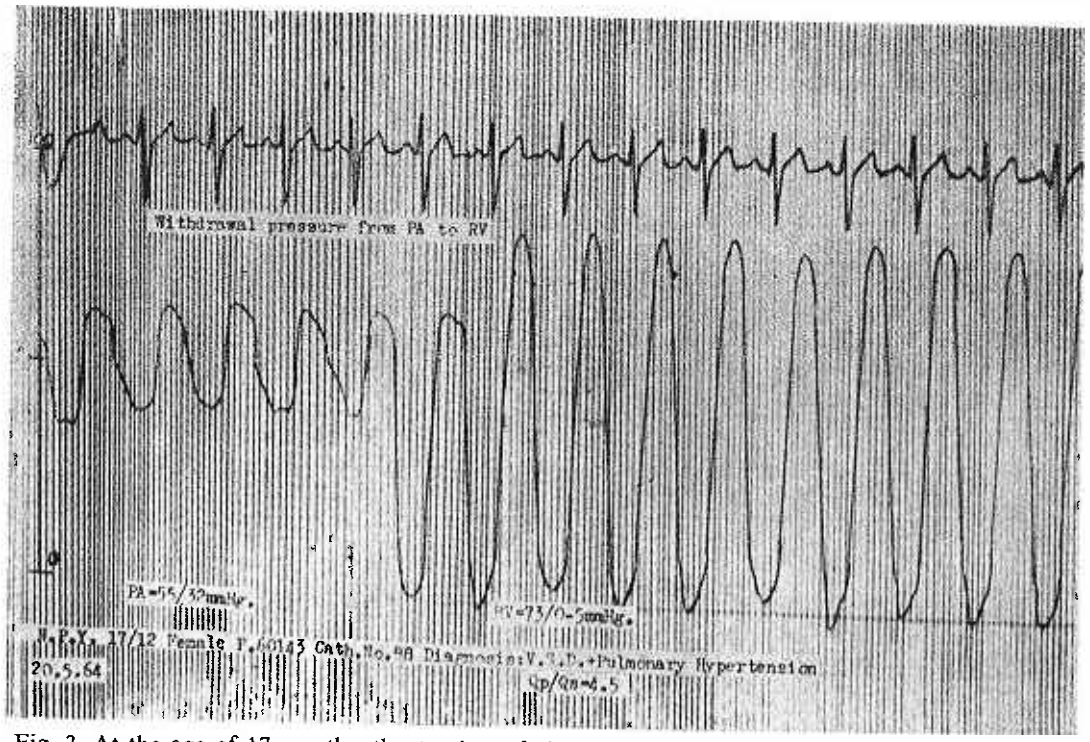


Fig. 3. At the age of 17 months, the tracing of the withdrawal pressure from pulmonary artery to right ventricle shows the pulmonary artery pressure to be 55/32 mm. Hg. and the right ventricular pressure 73/5 mm. Hg.

Her chest X-rays demonstrated a striking change from marked pulmonary plethora to oligaemic lung fields as shown in Fig. 2.

She was also re-investigated with a right heart study and right ventriculogram $3\frac{1}{2}$ years later and the haemodynamic data are as follows (Table I).

Withdrawal pressure tracings demonstrated clearly a systolic gradient of 73 mm. Hg. across the infundibulur as shown in Figs. 3 and 4.

Right ventriculogram (19.5.67) showed infundibular stenosis with a right-to-left shunt at the ventricular level consistent with Fallot's Tetralogy (Fig. 5).

Case No. 2. Z. bte S. 2 years female. P. No. 86116

Her birth history was unremarkable. She was first referred at the age of 2 months because of a cardiac murmur. She had recurrent attacks of bronchitis adequately treated without hospitalization. There was no history of cyanosis or cardiac failure. Clinically she was diagnosed as a case of ventricular septal defect with pulmonary hypertension. This was confirmed by cardiac catheterization at the age of 1 year.

She was subsequently followed-up in the Cardiac Clinic. She progressed satisfactorily and remained asymptomatic after $1\frac{1}{2}$ years of age. On examination, at 2 years of age, she still had no cyanosis or clubbing. Her heart was not clinically enlarged. She had a systolic thrill at the 3rd left intercostal space at the sternal edge and an ejection systolic murmur with a soft pulmonary second sound, Hb.=11.0 gm.%, PCV=34%.

Her serial electrocardiographic changes are as shown in Fig. 6.

The mean QRS axis from +45° at 13 months, changed to +135° at 2 years, and progressive

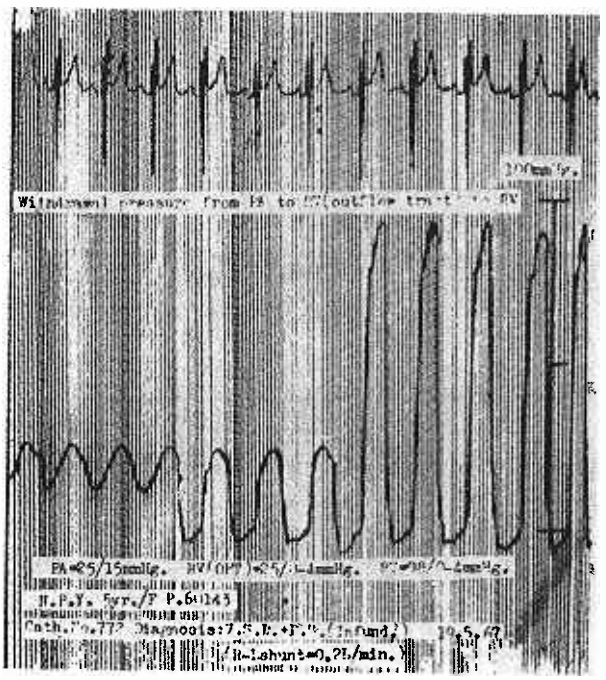


Fig. 4. At a repeat cardiac catheterization done at the age of 5 years, the withdrawal pressure tracing shows an acquired infundibular obstruction with a systolic gradient of 73 mm. Hg.

TABLE I

Catheter No. 88 (20-5-64)

Pressure
- PA wedged mean 10 mm. Hg.
- PA 55/32 (39) mm. Hg.
- RV outflow tract
- RV 73/0-5 mm. Hg.
- RA mean 5 mm. Hg.
- FA 100/60 (73) mm. Hg.

Oxygen content
- PA 15.0 Vol.%
- RV 15.5 Vol.%
- RA 11.5 Vol.%
- FA 16.0 Vol.%

Capacity 16.5 Vol.%

Catheter No. 772 (19-5-67)

Pressure
- PA wedged mean 10 mm. Hg.
- PA 25/15 (18) mm. Hg.
- RV outflow tract 25/0-4 mm. Hg.
- RV 98/0-4 mm. Hg.
- RA mean 4 mm. Hg.
- FA 95/80 (85) mm. Hg.

- PA 13.0 Vol.% (72%)
- RV 13.0 Vol.% (72%)
- RA 12.5 Vol.% (70%)
- FA 16.0 Vol.% (90%) (crying)
- BA 17.0 Vol.% (94%) (at rest)

Capacity 18.0 Vol.%

	Systemic	Pulmonary	Systemic	Pulmonary
Cardiac Output	1.9 L/min.	8.3 L/min.	2.6 L/min.	2.4 L/min.
Cardiac Index	4.4 L/min./m.^{2}	19.8 L/min./m.^{2}	4.4 L/min./m.^{2}	3.9 L/min./m.^{2}
Qp/Qs	4.5	Qp/Qs	0.9	
Rp	1.5 units	Rp	2.0 units	
Pp/Ps	0.75	Pp/Ps	0.26	

(Qp/Qs = Pulmonary to Systemic flow; Rp = Pulmonary Vascular resistance; Pp/Ps = Pulmonary to Systemic systolic pressure ratio)

TABLE II

Catheter No. 576 (29-6-66)

Pressure mm. Hg.
- PA wedged mean 10 mm. Hg.
- PA 52/24 (33) mm. Hg.
- RV outflow tract
- RV 57/0-3 mm. Hg.
- RA mean 4 mm. Hg.
- BP 100/70 mm. Hg.

Oxygen content Vol.%
- PA 11.0 Vol.% (89%)
- RV 11.0 Vol.% (85%)
- RA 7.0 Vol.% (70%)
- FA 13.0 Vol.% (98%)
- Capacity 13.5 Vol.%

Catheter No. 802 (29-6-67)

Pressure mm. Hg.
- PA wedged mean 8 mm. Hg.
- PA 20/8 (12) mm. Hg.
- RV outflow tract 20/0-3 mm. Hg.
- RV 87/0-3 mm. Hg.
- RA mean 4 mm. Hg.
- FA 89/60 (70) mm. Hg.

Oxygen content Vol.%
- PA 8.0 Vol.% (65%)
- RV 8.0 Vol.% (70%)
- RA 7.5 Vol.% (62%)
- FA 12.5 Vol.% (99%)
- Capacity 12.5 Vol.%

	Systemic	Pulmonary	Systemic	Pulmonary
Cardiac Output	1.0 L/min.	2.3 L/min.	2.0 L/min.	2.0 L/min.
Cardiac Index	3.3 L/min./m.^{2}	7.9 L/min./m.^{2}	4.4 L/min./m.^{2}	4.4 L/min./m.^{2}
Qp/Qs	2.4	Qp/Qs	1.0	
Rp	3.5 units	Rp	0.9 units	
Pp/Ps	0.52	Pp/Ps	0.22	

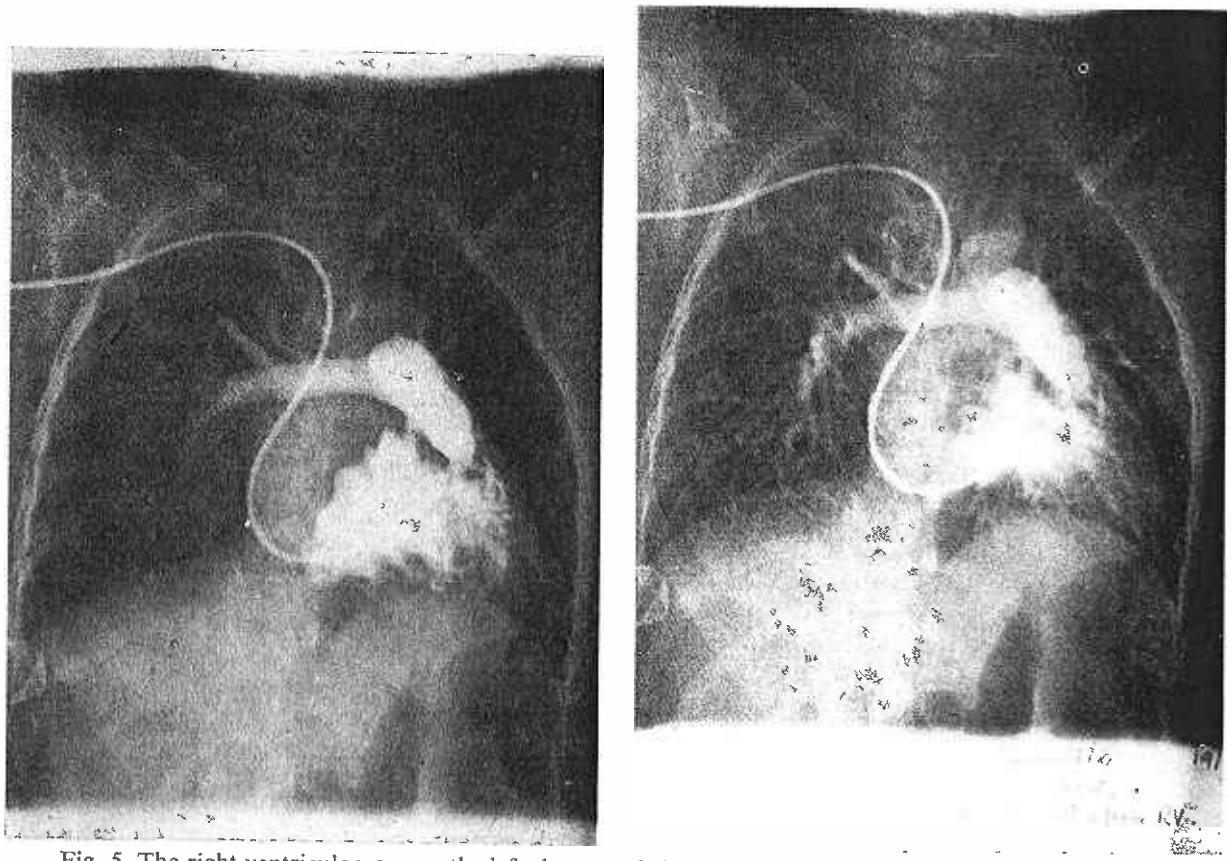


Fig. 5. The right ventriculogram on the left shows an infundibular stenosis, and that on the right shows opacification of the aorta as a result of right-to-left shunt across the ventricular septal defect.

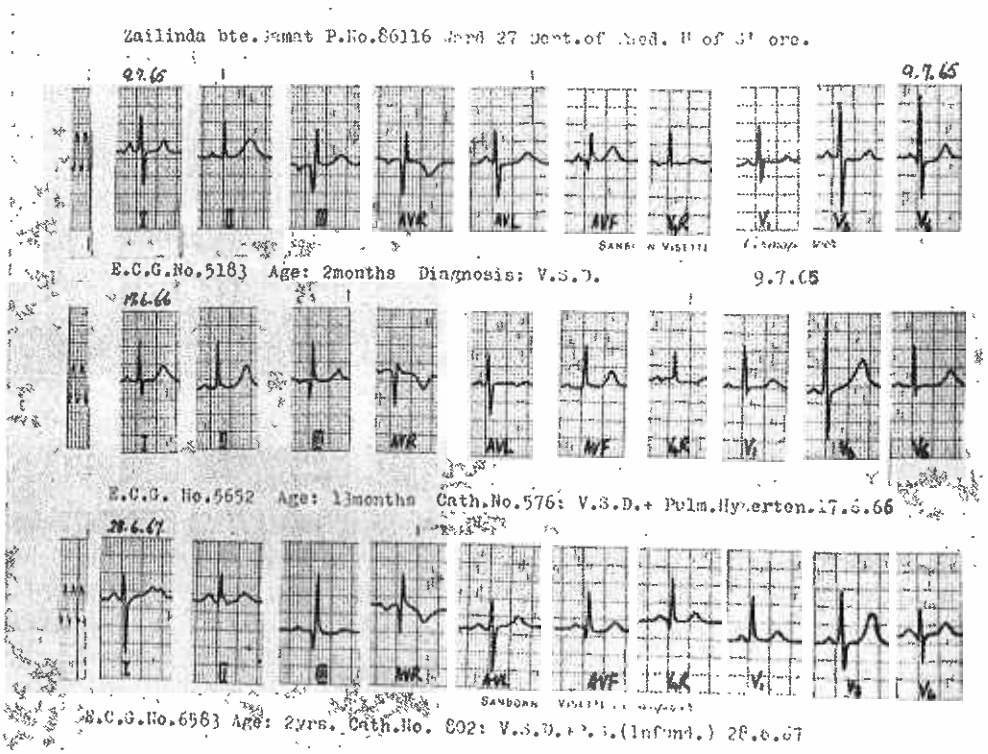


Fig. 6. Serial electrocardiograms done at 2 months, 13 months and 2 years of age, show the increase in right ventricular hypertrophy.

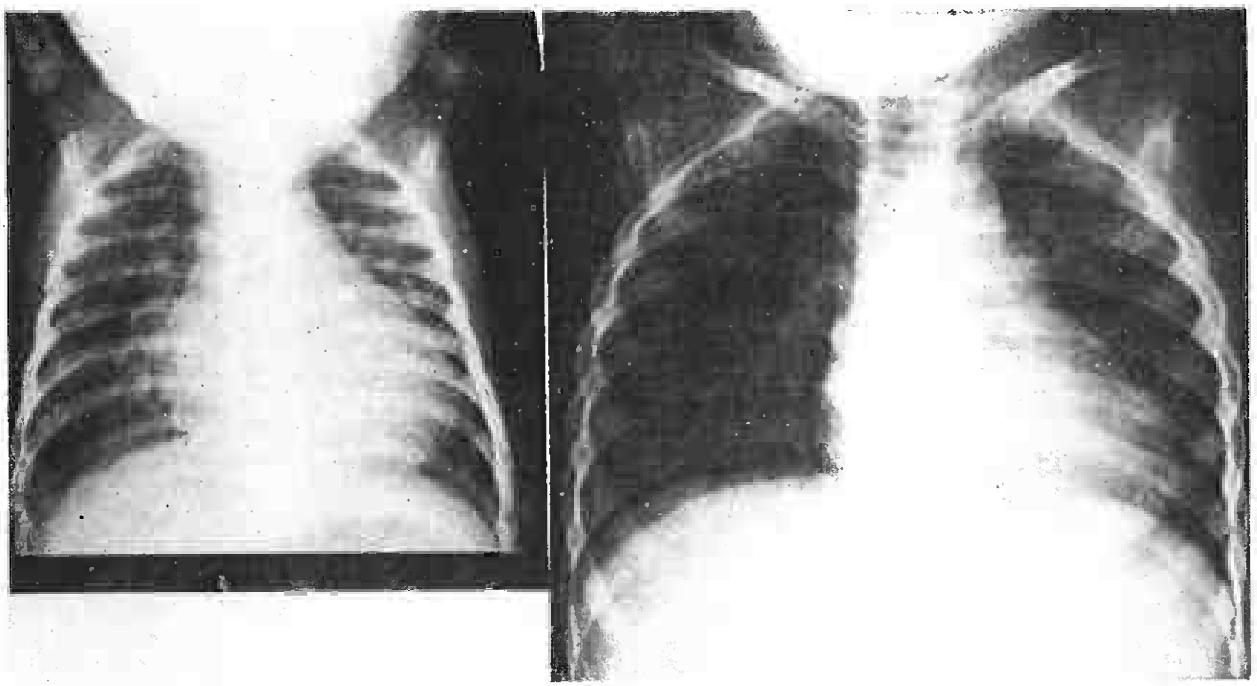


Fig. 7. The chest X-ray on the left shows marked pulmonary plethora and that on the right, normal lung fields.

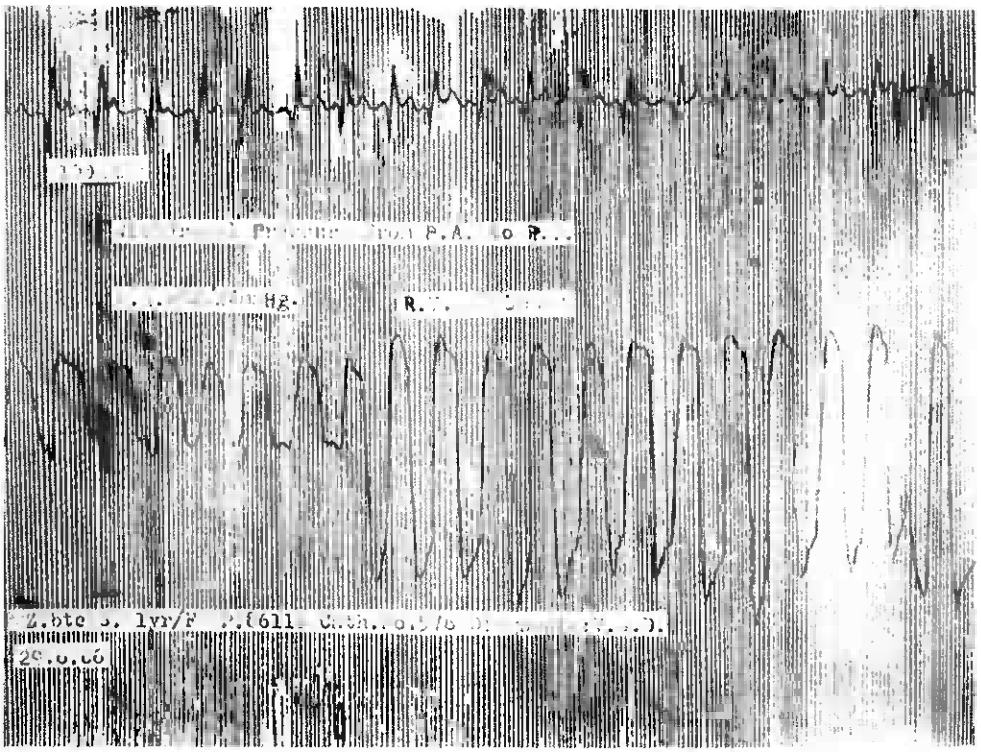


Fig. 8. Shows the pressure recording from the pulmonary artery to the right ventricle at the age of 1 year. The pulmonary artery pressure was 52/24 mm. Hg. and the right ventricular pressure 57/3 mm. Hg.

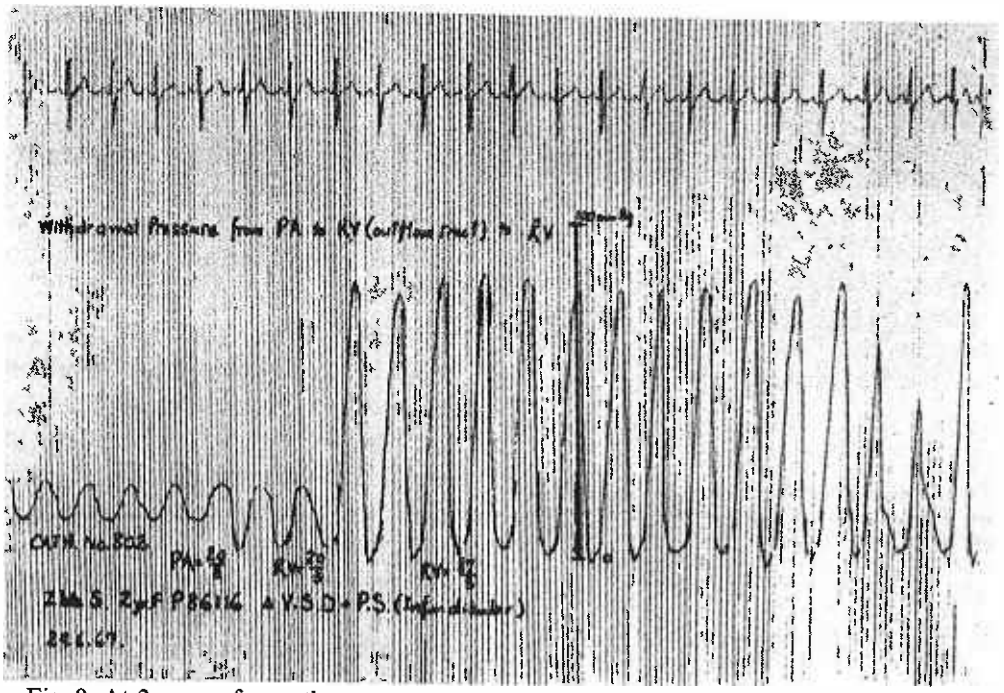


Fig. 9. At 2 years of age, the pressure recording shows an acquired infundibular stenosis with a systolic gradient of 67 mm. Hg.

increase in right ventricular hypertrophy was also demonstrated.

Her chest radiographic appearance changed from one with marked pulmonary plethora to one with normal lung fields as shown in Fig. 7.

Her haemodynamic data are as follows (Table II).

Comparison of the tracings of the withdrawal pressure from pulmonary artery to right ventricle in the two cardiac catheterizations is shown in Figs. 8 and 9.

The tracings of the withdrawal pressure clearly demonstrated the acquired gradient of 67 mm. Hg. across the infundibulum over a period of 12 months.

This patient had a ventricular septal defect associated initially with moderate pulmonary hypertension and a moderate L-R shunt confirmed by cardiac catheterization at the age of 1 year. Gradually, she developed pulmonary infundibular obstruction which rendered her asymptomatic because of a balanced pressure between the right and left ventricles and no L-R shunt was detectable by our present technique. This change was also confirmed by serial electrocardiograms and chest-X-rays.

Case No. 3. T.S.P. $2\frac{1}{2}$ years male. P. No. 82910

He was the youngest of 10 siblings. Birth history was normal and his birth weight was 6 lbs. 10. oz.

The cardiac lesion was first detected when he was admitted with chest infection and cardiac failure at the age of 2 months. His subsequent admissions were at three months, eleven months, 1 year 4 months and 2 years, all for repeated chest infection and cardiac failure. He failed to thrive, was a difficult feeder, had marked nutritional anaemia and his effort tolerance was poor.

Cardiac catheterization was first done on him at the age of eleven months and the diagnosis of ventricular septal defect with pulmonary hypertension was confirmed. He was closely observed in the Cardiac Clinic at monthly intervals and gradual improvement was seen after the age of $1\frac{1}{2}$ years.

Slight cyanosis was noted for the first time at the age of $2\frac{1}{2}$ years. At this time although symptomatically he had improved to some extent, he was still very stunted in growth. He had a marked precordial bulge with a systolic thrill and an ejection systolic murmur Grade 3/6 at the 3rd left, intercostal space at the sternal edge. The pulmonary closure sound was diminished. His pulse was regular and of normal volume.

His serial electrocardiograms showed increasing right ventricular overload pattern (Fig. 10).

A second cardiac catheterization was done at $2\frac{1}{2}$ years of age and the withdrawal pressure from pulmonary artery to right ventricle showed a systolic gradient of 50 mm. Hg. at the infundi-

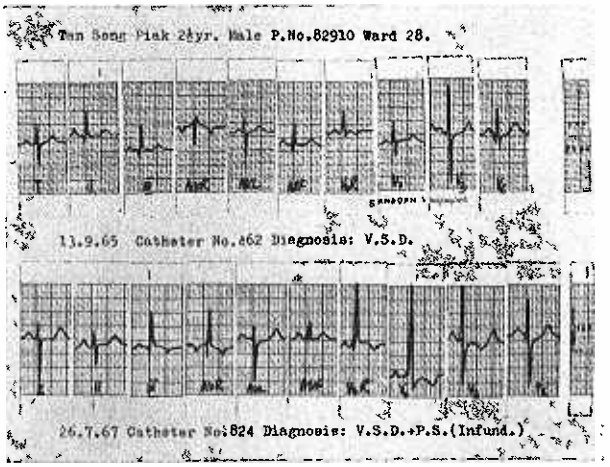


Fig. 10. Two electrocardiograms done with an interval of 22 months between them, show a progressive increase in right ventricular hypertrophy.

bular level. His haemodynamic data are as follows (Table III).

Comparison of withdrawal pressure tracings in both studies revealed an increase in infundibular gradient as shown in Figs. 11 and 12.

This patient has acquired over a period o eighteen months an infundibular obstruction severe enough to produce a balanced pressure between the two ventricles. This caused a drastic reduction of L-R shunt at the ventricular level, even producing a slight right-to-left shunt.

DISCUSSION

Gasul *et al* (1957)[1] described the new concept of the natural transformation of ventricular septal defect into one with an acquired infundibular obstruction severe enough to give rise to a cyanotic or non-cyanotic type of Fallot's tetralogy. Hypertrophy of the crista supraventricularis and perhaps, also of its septal or parietal band, occurs under the prolonged stress of torrential pulmonary blood and high pulmonary artery pressure. When the hypertrophy is severe enough to create an obstruction at the right ventricular outflow tract, the patient seems to improve spontaneously because the obstruction helps to protect the pulmonary vascular bed by reducing the left-to-right shunt at the defect level. The infundibular hypertrophy with subsequent outflow tract obstruction has been regarded as an adaptation to protect the pulmonary vascular bed against the excessive pulmonary blood flow as suggested by Watson *et al* (1965)[8]. This combination of ventricular septal defect and reactive infundibular stenosis represents an acyanotic type of tetralogy of Fallot.

Lucas *et al* (1962)[5] reported that anomalous muscular bundles in the right ventricle may accompany a ventricular septal defect, giving rise to functional obstruction at the inflow portion of the right ventricle just beyond the level of the septal defect. Depending on the severity, the resulting functional and clinical picture is similar to that of acyanotic or cyanotic tetralogy of Fallot.

To demonstrate this change of events in the natural history of ventricular septal defect, serial cardiac catheterization is necessary and should be carried out during the first few years of life when the haemodynamic changes are most prominent. In this present series, three patients demonstrating the acquired infundibular obstruction are described. Case one shows quite remarkable changes from birth to the age of five years and these are confirmed by serial haemodynamic studies and selective angiocardiography. Cases 2 and 3 also show the same changes, but selective angiocardiography was deferred because a repeat study is contemplated only when they are ready for open heart repair.

The incidence of this natural transformation in the mild form is certainly common but the incidence of acquired infundibular obstruction of the severity seen in the above cases is not known. Walker *et al* (1965)[7] in their analysis of 415 catheterised cases of which 90 serial haemodynamic studies, found that there was a frequent association (11%) between ventricular septal defect and right ventricular outflow tract obstruction. Some patients developed increasing right ventricular outflow tract obstruction during study.

Held *et al* (1963)[2] confirmed by serial roentgenography the progressive increase in the systolic pressure gradient across the right ventricular outflow tract in the evolution of infundibular hypertrophy and outflow tract stenosis. In a group of nine patients followed for three to eleven years, the roentgenographic changes included a diminution in the peripheral pulmonary vasculature, diminution in the size of the main pulmonary artery, reduction in the size of the left atrium, decrease of the degree of cardiomegaly and an increase in the size of the aorta. There is no known method of predicting when and in which type of ventricular septal defect it is going to occur. From the evidence available, if it does occur, it will be during the first few years of life.

Since mortality is highest in patients with ventricular septal defect during infancy, the

suggestion of creating this situation artificially by 'banding' the pulmonary artery was made by Muller and Dammann (1952).[6] They based their technique on the experimental work of Hufnagel *et al* (1951)[3] and reported that, by the artificial production of pulmonary obstruction, they could relieve the overload of the pulmonary circulation from a large left-to-right shunt.

'Banding' of the pulmonary artery is a surgical procedure designed to simulate the naturally occurring pulmonary obstruction which reduces the left-to-right shunt at the ventricular septal defect, thus protecting the pulmonary vascular bed. This procedure has been improvised to salvage patients with a large defect and torrential left-to-right shunt, and also patients in whom open heart closure of the defect carries high mortality risks such as during infancy and the early years of life. It is a palliative procedure and is a recognised form of treatment.

'Banding' of the pulmonary artery is not without risks. Since this procedure has been adopted, increasing incidence of complications is being reported. Perhaps, if it were possible to predict in which patients naturally acquired infundibular obstruction would occur, unnecessary 'banding' could be avoided. Therefore, the study of this problem should continue, in the hope that further light can be thrown on the role acquired infundibular obstruction plays in the natural history of ventricular septal defect.

TABLE III

Catheter No. 462 (27-11-65)

Pressure mm. Hg.

PA wedged mean 9 mm. Hg.
PA 54/18 (30) mm. Hg.
RV outflow tract
RV 85/0-4 mm. Hg.
RA mean 7 mm. Hg.
BP 110/50 mm. Hg.

Oxygen saturation % (content Vol. %)

PA 89% (10.5 Vol. %)
RV 88% (10.5 Vol. %)
RA 74% (9.0 Vol. %)
FA 99% (12.0 Vol. %)

Catheter No. 825 (28-7-67)

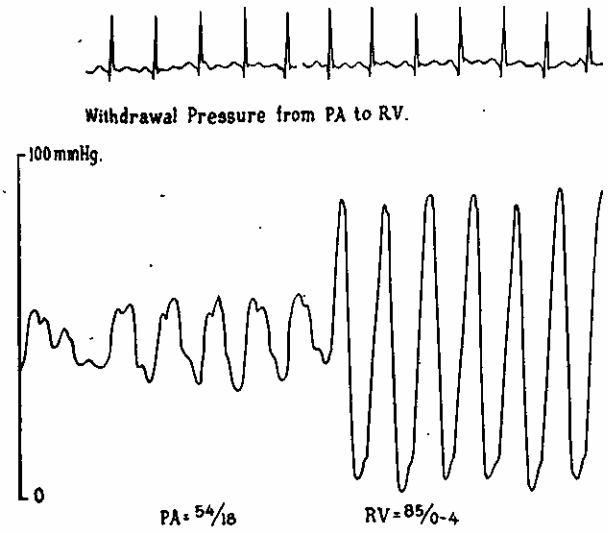
Pressure mm. Hg.

LA mean 8 (through PFO) mm. Hg.
PA 42/16 (25) mm. Hg.
RV outflow tract 42/0-6 mm. Hg.
RV 92/0-7 mm. Hg.
RA mean 6 mm. Hg.
FA 90/61 (71) mm. Hg.

Oxygen saturation % (content Vol. %)

PA 87% (11.0 Vol. %)
RV 85% (11.0 Vol. %)
RA 81% (10.0 Vol. %)
FA 93% (12.5 Vol. %)

	Systemic	Pulmonary	Systemic	Pulmonary
Cardiac Output	2.2 L/min.	4.5 L/min.	2.0 L/min.	2.6 L/min.
Cardiac Index	6.6 L/min./m.^{2}	13.2 L/min./m.^{2}	5.0 L/min./m.^{2}	6.6 L./min./m.^{2}
Qp/Qs	2.0	Qp/Qs	1.3	
Rp	1.6 units	Rp	2.6 units	



T.S.P. 11/12 male P No. 82910 Catheter No. 462 Δ V.S.D. 27-1-1966.

Fig. 11. The pressure recording shows the pulmonary artery pressure to be 54/18 mm. Hg. and the right ventricular pressure 85/4 mm. Hg. at the first cardiac catheterization.

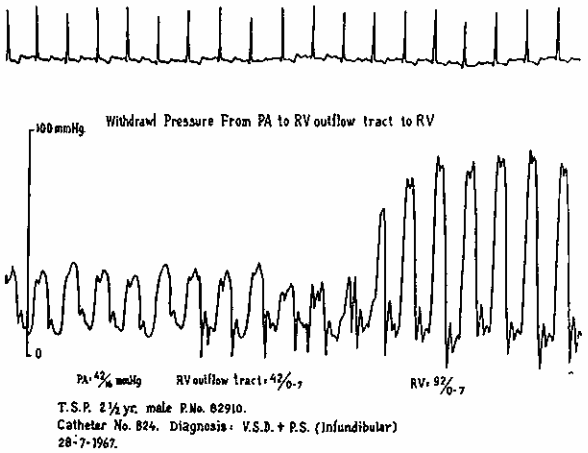


Fig. 12. The withdrawal tracing across the right ventricular outflow tract done at the second catheterization shows the systolic gradient to be 50 mm. Hg., and also demonstrates an acquired obstruction at the infundibular level.

SUMMARY

Three patients with naturally acquired infundibular obstruction in ventricular septal defect with pulmonary hypertension are reported. Two of the patients had severe obstruction causing a right-to-left shunt and subsequent cyanosis simulating Fallot's tetralogy, but in the remaining one, the pressures in both ventricles were balanced and no shunt could be detected at all by our present technique. All observations were confirmed by serial cardiac catheterization. The significance of this occurrence is discussed.

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