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Acquired pulmonary atresia in tetralogy of Fallot with a functioning Blalock-Taussig shunt

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Patients have been described who, presenting clinically as cases of the tetralogy of Fallot, have yet been found, on further study, to have pulmonary atresia. There have been others who were thought to have had flow across the pulmonary valve at the time of the performance of a systemic to pulmonary artery shunt but in whom the pulmonary valve was later shown to be atretic (Taussig, 1965). This finding has been interpreted as evidence of probable acquired atresia of the pulmonary valve possibly through some form of late fusion of the cusps (Edwards, 1965). Sabiston, Cornell, Criley, Neill, Ross, and Bahnson (1964) have presented excellent evidence that this can indeed occur. In this report a patient is presented in whom incontrovertible radiographic proof of pulmonary valve patency was obtained at catheterization yet complete atresia was found at operation six months later.

CASE REPORT

When first seen, the patient was 2 years old with a history of cyanosis since infancy, squatting, increasingly frequent and severe spells of unconsciousness, and dyspnoea on exertion. On examination she had central cyanosis, clubbing, an ejection systolic murmur maximal in the second left interspace, a single second sound, and electrocardiographic evidence of right ventricular hypertrophy. Because of the worsening symptoms a left subclavian to pulmonary artery anastomosis was made. After a stormy post-operative course the child did very well. At the age of 8 years some increase in cyanosis was noted and she was catheterized with a view to corrective surgery. The physical findings at this time continued to be typical for tetralogy of Fallot with a functioning Blalock anastomosis. The findings at cardiac catheterization confirmed the clinical impression. During the examination a catheter (no. 5 Lehman) was passed through the pulmonary valve into the artery. An injection of angiographic medium outlined the artery and valve well and confirmed the position of the catheter. During the next six months before surgery the patient's exercise tolerance decreased and she became more cyanosed. The physical findings were otherwise unchanged.

At operation, when the subclavian to pulmonary artery shunt was momentarily occluded, both the systolic and the diastolic pulmonary artery thrills disappeared. The main pulmonary artery was quite adequate in size, being about half the diameter of the aorta. After cardiopulmonary bypass had been started and the right ventricle opened, very extensive muscular infundibular obstruction was found. The infundibular chamber was small but the pulmonary valve, although recognizable as probably a once tricuspid structure, was completely blocked (Fig. 1). No opening could be found on careful examination with a fine probe. Cuts were made through the valve tissue at the sites of probable commissural fusion, and bronchial collateral blood flow immediately came back from the pulmonary artery. A 1·2 cm. diameter dilator was easily passed through the valve and, although an outflow tract patch was used because of the narrowness of the infundibular chamber, it was not necessary for this to cross the valve ring. Her convalescence was marred by a wound infection but was haemodynamically uneventful. The patient did not experience the temporary period of congestive heart failure commonly seen after the complete repair of tetralogy of Fallot. From the first post-operative day a loud pulmonary regurgitant murmur was heard.

DISCUSSION

This case documents the closure of a previously patent pulmonary valve in a patient with tetralogy of Fallot and a functioning Blalock anastomosis. The closure took place during a six-month period and was accompanied by clinical deterioration, which was thought pre-operatively to be related to relative narrowing of the Blalock anastomosis with growth of the patient, but was obviously due, in part at least, to the stopping of what little blood flow there had been from the right ventricle.

In speculating on the cause of this phenomenon, it is possible that with marked infundibular

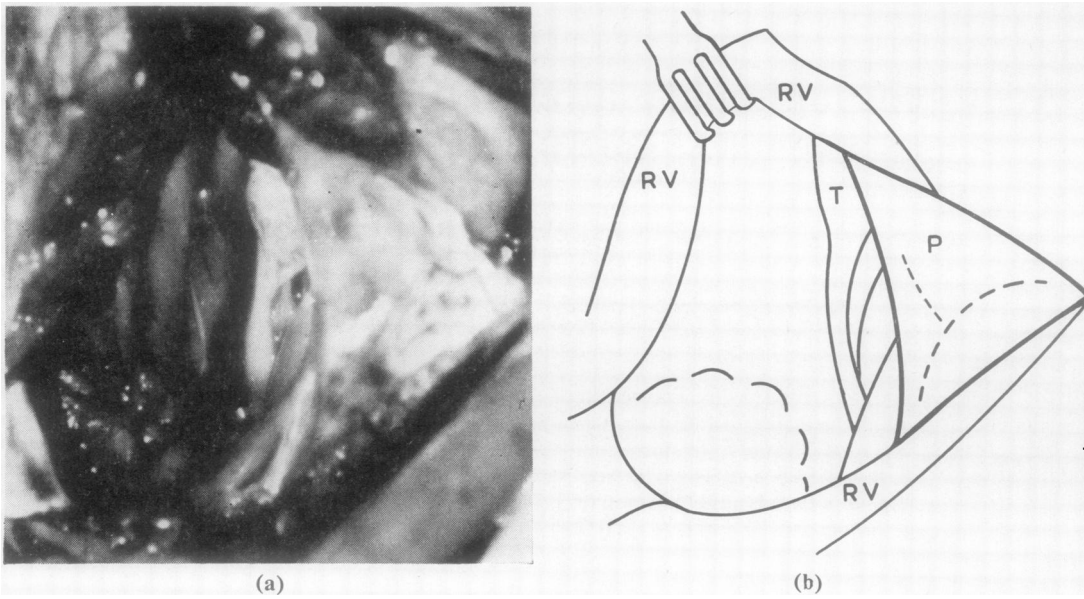

FIG. 1. (a) *Photograph of the pulmonary valve seen through the right ventriculotomy;* (b) *tracing of the photograph. RV, right ventricular wall; T, tricuspid valve hiding edge of ventricular septal defect; P, pulmonary valve showing the lines of fusion of the cusps.*

stenosis allowing only a small part of the right ventricular output to enter the pulmonary arteries in the usual way and a good systemic flow entering it distally, the valve would have a very limited range of movement with each cardiac cycle and would, for the most part, be closed. The range of movement of these cusps might, in any case, have been limited by partial congenital fusion of the commissures. In the presence of relatively stagnant blood, fibrin deposition could occur on the cusps and result ultimately in their adhesion. Another possibility is that the passage of the catheter through the valve damaged the cusps, leaving raw areas on their apposing surfaces which later became stuck to each other. It is, of course, unknown whether some fusion of the cusps had been taking place before the catheterization. The possibly limited movement of the cusps would aggravate the tendency for injury to result in their adhesion. The presence of this valve deformity did not affect the operative procedure adversely in any way other than to make post-operative pulmonary incompetency more likely than usual.

It is of interest that, in discussing this case with other cardiac surgeons, it became apparent that similar cases had been seen quite often although not with documentation of previous patency.

SUMMARY

A case of tetralogy of Fallot is presented in which the pulmonary valve was observed to become completely occluded in the presence of a functioning Blalock-Taussig anastomosis. This event probably occurs commonly enough for every cardiac surgeon to encounter it from time to time. It need not adversely affect the results of complete correction.

REFERENCES

Edwards, J. E. (1965). Personal communication.

Sabiston, D. C., Jr., Cornell, W. P., Criley, J. M., Neill, C. A., Ross, R. S., and Bahnson, H. T. (1964). The diagnosis and surgical correction of total obstruction of the right ventricle. *J. thorac. cardiovasc. Surg.*, **48**, 577.

Taussig, M. B. (1965). Personal communication.