Atrial Septal Defect Complicated by Pulmonary Hypertension—A Long Term Follow Up

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PULMONARY hypertension in atrial septal defect is serious and warns of impending difficulty in untreated cases [4] and is responsible for a disturbing increase in surgical mortality.[1, 2, 5] Our own experience reveals a ten-fold increase in surgical mortality in the pulmonary hypertensive group.

The increased risk of operation and the uncertain benefit to the survivors has prompted the study reported here.

Material

At the University of Western Ontario Hospitals 72 atrial septal defects have been repaired surgically since 1956. Twelve of these patients had pulmonary hypertension as defined by pulmonary artery pressures greater than 50 mm. of mercury systolic.

There was a strong predominance of women: 10 to 2 men. Ages varied from 33 to 62 years (average 40) and the pulmonary artery pressures varied from 56 to 105 mm. of mercury with an average of 75.

All patients were in Class 3 or Class 4 of the New York Heart Association classification.

Right heart catheter studies revealed a left-to-right shunt still evident in all patients. Pulmonary-to-systemic flow ratios varied from 1.5 to 1 to 3 to 1, and the arterial oxygen saturations varied from 84 to 100%. There was no clinical evidence of cyanosis at rest.

Nine of the atrial septal defects were of the ostium secundum variety and 3 were of the sinus venosus variety. The defects were repaired by direct suture or with a patch of Dacron, Teflon or autogenous pericardium.

Results

Two patients died in the postoperative period, one of acute myocardial infarction and the other from irreversible right heart failure. The 10 survivors were subjected to repeated clinical and hemodynamic studies. Follow-up periods varied from 2½ to 10 years.

The postoperative complication rate was high due to arrythmia in four, resistant right heart failure in four and a non-fatal air embolism in one. All patients were subjected to tracheostomy and eight were ventilated with high pressures (up to 35 mm. Hg) to reduce the inevitable increase in pulmonary vascular resistance that occurs in the first 48 hours after operation.[3]

Of the 10 survivors (who were Class 3 preoperatively), three now lead an unrestricted life (Class 1); six are in Class 2 and are able to do their housework with occasional need for additional rest, one patient is recategorized in Class 3 because of development of angina that is probably unrelated to the atrial septal defect.

Presented at the Annual Meeting of the American Surgical Association, May 11–13, 1967, Colorado Springs, Colorado.

The radiologic changes depend primarily on the degrees of pulmonary vascular resistance preoperatively. Those with moderate or severe elevation of the pulmonary vascular resistance have no demonstrable change radiologically. Those with slight or normal pulmonary vascular resistance show a reduction in heart size.

Pulmonary artery pressure is reduced in all survivors and falls within normal limits in five (50%). It is less than half of the preoperative value in 90% of patients.

In one patient (a 46-year-old woman) the pulmonary artery pressure (as measured at right heart catheter study 2 years after operation) increased from 60 to 68 mm. Hg systolic. Reoperation (because of a residual left to right shunt) was carried out and a subsequent catheter study revealed a pulmonary artery pressure of 38 mm. Hg.

Changes in pulmonary vascular resistance following operation are more subtle than those in pulmonary artery pressure and although the former tend to recede postoperatively, they remained grossly elevated in three (30%); 5.6, 3.1 and 2.8 Paul Wood, respectively. In general these patients had a fall in resistance to two-thirds of the preoperative level but this did not occur in the first postoperative year.

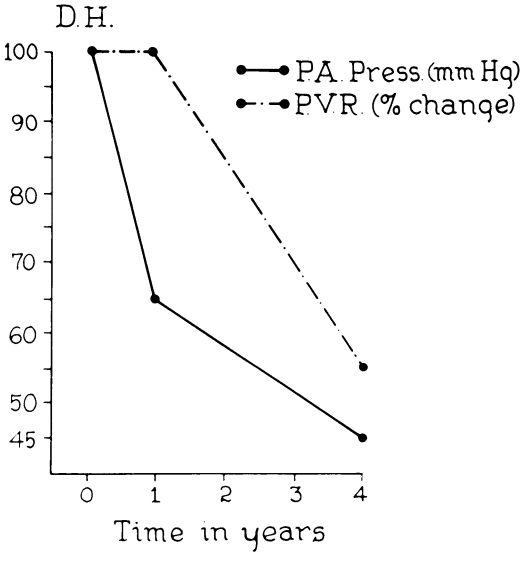

FIGURE 1.

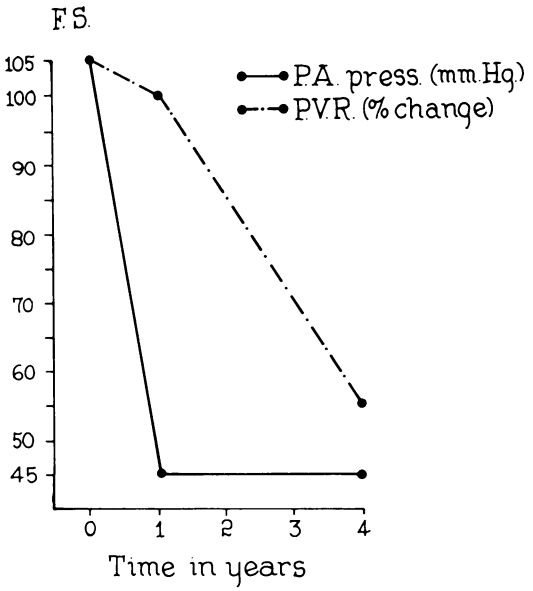

FIGURE 2.

Pulmonary hypertension in atrial septal defect is due to two factors—the hyperkinetic or flow factor and the factor of pulmonary vascular resistance. Pulmonary hypertension is almost unknown in children with atrial septal defect who live at or near sea level and occurs in only approximately 20% to 30% of adults.

Increase in pulmonary resistance may be due to organic changes in the pulmonary artery or pulmonary arteriole and is manifest by medial hypertrophy or intimal hyperplasia or fibrosis. The hyperkinetic factor is abolished with closure of the left-to-right shunt and is believed responsible for the early reduction in the pulmonary artery pressure following operation.

Of particular interest are two patients with severe pulmonary hypertension (Fig. 1). The pulmonary pressure of 100 mm. of mercury fell to 65 mm. of mercury one year following operation; there was no change in the pulmonary vascular resistance at this time. A hemodynamic study 3

years later revealed a fall in the pulmonary vascular resistance to 55% of the preoperative level. This was associated with a further fall in the pulmonary artery pressure to 45 mm. of mercury.

The second patient demonstrates a slight reduction in pulmonary vascular resistance one year following operation at the time of a marked fall in the pulmonary artery pressure (to 45 mm. of mercury (Fig. 2). A study 3 years later revealed a further change in pulmonary vascular resistance to 55% of the preoperative level with no further reduction in pulmonary artery pressure. The cardiac output in this patient improved 50% in this interval.

Summary

Ten patients with pulmonary hypertension survived closure of artrial septal defects and were subjected to repeated clinical and hemodynamic assessment. Clinical improvement was noted in all as was a fall in pulmonary artery pressure. Electrocardiogram was moderately improved, x-ray varied from no change to moderate improvement and the pulmonary vascular resistance showed a late regression to approximately two-thirds of the preoperative value in three patients with severe pulmonary hypertension.

We conclude that pulmonary hypertension is not a contraindication to operation in atrial septal defect. There is long-term benefit to these patients and there is evidence from this small series to suggest a recession in pulmonary vascular resistance in the late follow-up period, even allowing for the possible errors in the measurement of this parameter.

Bibliography

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Discussion

Dr. Donald J. Ferguson (Chicago): Underlying the clinical phenomena which have been described by Dr. Coles are the remarkable reactions of the pulmonary arteries to the stress of hypertension. Dr. James Esterly and I have studied by electron microscopy serial lung biopsies from dogs in which pulmonary hypertension was produced by a left-to-right shunt.

(Slide) This is an electron micrograph of an endothelial cell from a normal small pulmonary artery in the dog. The particular character of this cell to which I wish to call your attention is the lack of very much visible organization in the cytoplasm. There are only a few mitochondria visible.

(Slide) The next slide shows a similar cell from a dog which has been subjected to pulmonary hypertension for a few weeks. This is a characteristic appearance of the endothelial cells, and the earliest change that we have been able to see in response to this stress. The changes here are the presence of numerous mitochondria, a rough endoplasmic reticulum, and small groups of ribosomes in the cytoplasm. There is now a characteristically enlarged Golgi apparatus present. Eventually these endothelial cells undergo degeneration, and then they all vanish, and the further process of arteriosclerosis goes on. So that to speak of the intimal lesion as intimal proliferation is incorrect.

(Slide) This shows another phenomenon which goes on after about 2 months of hypertension, and that is a migration of smooth muscle cells through the internal elastic lamina to fill up what was previously an intimal space, and eventually to occlude the lumen of some of the vessels. These cells are identifiable as smooth muscle by the presence in them of myofibrils, by numerous