CLINICOPATHOLOGIC CORRELATIONS

Pulmonary Vascular Disease Complicating the Blalock-Taussig Anastomosis

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SUMMARY The pulmonary vascular bed was evaluated following a Blalock-Taussig shunt performed in 36 patients for decreased pulmonary blood flow. The time of follow-up ranged from one to 21 years after operation. No patients developed severe pulmonary changes in less than eight years after institution of the shunt. Ten of 20 patients having a shunt eight years or longer developed some degree of pulmonary vascular disease, mainly in the form of intimal fibrosis. This was severe in six of the 20 patients. As the duration of the shunt increased, so did the incidence of pulmonary vascular disease.

ALTHOUGH THE BLALOCK-TAUSSIG SHUNT has been performed since 1945,1 the long-term effects upon the pulmonary vascular bed have been recorded only incompletely. While pulmonary vascular disease following the Waterston or Potts operations has been reported to occur in from 10 to 50% of cases, its incidence after the Blalock-Taussig shunt has been thought to be much lower. Daoud,2 Hancock,3 and Roberts4 and their associates have reported cases with hypertensive pulmonary vascular disease as a consequence of the Blalock-Taussig anastomosis. Ross and associates5 found only 11 cases of pulmonary vascular disease following the Blalock-Taussig operation performed in 224 cases.

This communication presents the histologic findings of the pulmonary arterial system in patients following the Blalock-Taussig shunt operation. Histologic findings were correlated with the duration of the shunt and the cause of death.

Procedure

Thirty-six patients from the University of Minnesota or Johns Hopkins Hospitals comprised the basis for this study. In each patient, a Blalock-Taussig shunt was created at some time between 1947 and 1964 for some form of pulmonary stenosis. In eight cases this operation was performed twice.

Of the 36 patients, 23 had the tetralogy of Fallot and four had transposition of the great vessels with a ventricular septal defect and pulmonary stenosis. The remaining patients had a variety of anatomic conditions, including pulmonary stenosis associated with cor triloculare, double outlet right ventricle or tricuspid stenosis. In three of the cases the ventricular septum was intact. The age of the patients ranged from six months to 17 years at the time of their initial shunt operation.

Eight patients had two Blalock-Taussig shunt operations.

The time between the operations varied from two to 11 years. Each of the patients died. The period between the time of death and initial (or only) shunt varied from one to 21 years. Patients having more than one shunt were classified according to the date of the initial operation.

Corrective surgery was attempted in about two-thirds of the patients. Twenty-two patients died in the early postoperative period following a corrective operation. The causes of death varied.

At the time of the autopsy, varying numbers of samples of lung were obtained for histologic examination and stained with H & E. In some instances, stains for elastic tissue were obtained on duplicate sections. The sections were then examined by two of the investigators and graded according to the classification given below. At the time of histologic examination of the lungs, the examiners did not know the duration of the shunt.

Results

Changes, when present, usually affected the small muscular pulmonary arteries more than the large muscular arteries. If some medial hypertrophy was present without intimal disease, the designation grade 1 was applied (fig. 1a). Grades 2 and 3 were based primarily upon degrees of intimal proliferation. In these latter two grades (fig. 1b and c), the media varied considerably; in some instances it was thin while in others it was hypertrophied.

When the vessels were considered normal, they showed neither intimal fibrosis nor medial hypertrophy. In many instances without pulmonary vascular disease, medial atrophy appeared to be present. No cases showed plexiform lesions. In isolated instances fibrous septa crossed the lumens of small arteries. These septa were like those recognized as occurring in pulmonary stenosis.6 When present, such lesions by themselves were not classified as pulmonary vascular disease for the purposes of this study. Of the 16 cases with some degree of pulmonary vascular disease, 12 showed change of the same degree in both lungs, while four developed vascular disease only in the lung which had received the subclavian artery. In no case had a pulmonary artery been divided to receive a shunt.

There appeared to be a relationship between the duration of the shunt and the extent of pulmonary vascular disease.

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The shortest post shunt period in which grade 3 changes were observed was eight years.

Using this observation, the cases were arranged in two groups as follows: group I, in which the post shunt period varied from one to seven years and group II in which the post shunt period varied from eight to 22 years (fig. 2).

No patients in group I had grade 3 changes. Ten of the 16 cases in this group had no evidence of pulmonary vascular disease, while six showed grade 1 or grade 2 changes. In group II, ten of the 20 patients had some degree of pulmonary vascular disease as follows: grade 1 and grade 2, three cases and grade 3, six cases. The post shunt period of the six cases with grade 3 changes were eight, nine, nine, 15, 16, and 21 years, respectively. Nine patients with shunts in place for 11 or more years showed no pulmonary vascular disease.

Cause of death was then compared to the severity of pulmonary vascular disease. The majority of patients died following a corrective procedure. Only two of the 22 patients who died following a corrective procedure exhibited grade 3 changes. The other causes of death in order of decreasing
frequency were congestive heart failure, arrhythmia and subacute bacterial endocarditis. There was no specific association of pulmonary vascular disease with these causes of death.

Comment

With the introduction of the Blalock-Taussig shunt operation, palliation was possible for a great number of conditions associated with pulmonary stenosis. One of the problems with the operation, however, was the need for a relatively large subclavian or pulmonary artery. In a small infant, a small subclavian artery makes the operation technically difficult or impossible. Subsequent experimentation led to the development of the Potts' and Waterston procedures.

The problems of small pulmonary or subclavian arteries are negated with these two procedures, since the descending or ascending aorta, respectively, is anastomosed to a pulmonary artery. Each of the latter operations has a greater applicability to the small infant than the Blalock-Taussig procedure. The Potts procedure, however, becomes complicated by a relatively high incidence of severe pulmonary vascular disease. Cole and associates found elevated pulmonary vascular resistance in 16 of 112 (14%) cases following the Potts procedure. Similarly, von Bernuth and associates reported 13 cases with pulmonary vascular disease among 26 patients following a Potts procedure (three to 12 years following operation). Cole and von Bernuth and their associates followed the Heath-Edwards classification of pulmonary vascular disease.

Although no accumulated data have yet been reported, the experience of Tay and associates suggests that pulmonary vascular disease of significant incidence may complicate the Waterston procedure.

While only six of their 36 total patients developed severe pulmonary vascular disease, the incidence tended to increase with the duration of the shunt, although not linearly. Roberts and associates reported four cases with Blalock-Taussig shunts of 13, 16, 17, and 19 years duration. Two showed no pulmonary vascular change, whereas another two had severe change.

Our results indicate that the Blalock-Taussig shunt carries little chance of complicating pulmonary vascular disease for at least seven years following performance of the procedure. None of our patients developed histologic evidence of severe pulmonary vascular disease during this time interval.

Finally, although no patients developed severe vascular changes during the first seven years postoperatively, 50% of the patients with a shunt longer than seven years had definite evidence of some degree of pulmonary vascular disease.

References

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