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Aneurysmal Degeneration of Blalock-Taussig Shunts: Identification and Surgical Treatment Options

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Many Blalock-Taussig shunts (subclavian to pulmonary artery anastomoses) have been created and a significant number are still being done. Two cases of aneurysmal degeneration of a Blalock-Taussig shunt and their management are described. Development of this rare complication may be related to large shunt flow and long duration. Large, symptomatic or enlarging aneurysms should be repaired and smaller ones studied by serial computed axial tomography. A simple and safe approach to correct this lesion is division and oversewing of the proximal subclavian artery through an anterior approach, assuming adequate pulmonary blood flow is already present or can be established concomitantly.

The subclavian to pulmonary artery anastomosis, or Blalock-Taussig shunt, has been utilized as a palliative procedure in children with cyanotic congenital heart disease since 1945 (1). Many shunts were created before the advent of complete surgical correction (2–4) and some patients are still living with their shunt (3). Despite advances in surgical techniques allowing for total correction of tetralogy of Fallot and other forms of cyanotic congenital heart disease in infancy (5–8), there are still significant numbers of patients receiving a Blalock-Taussig shunt (8–14).

Several early and late complications have been reported in association with the subclavian artery to pulmonary artery anastomosis (2–4,15–21). The formation of a subclavian artery aneurysm after a Blalock-Taussig shunt has only been reported once, and surgical correction was not recommended in that case (17). We subsequently operated on that patient and one other symptomatic patient with a Blalock-Taussig shunt aneurysm.

Case Reports

Case 1. A 37 year old woman with tetralogy of Fallot underwent creation of a right Blalock-Taussig shunt during 1947 when she was 5½ years old. Four years before admission, she had only mild exercise limitation and was found to have aneurysmal dilation of the subclavian portion of the shunt, discovered on a preemployment chest X-ray film (Fig. 1A). She was followed up with serial chest roentgenograms without appreciable increase in the mediastinal silhouette. Physical examination revealed an asymptomatic pulsatile mass at the base of the right side of the neck. There was no evidence of superior vena cava obstruction.

Cardiac catheterization revealed the classic findings of tetralogy of Fallot. A large aneurysm of the right subclavian artery was found just before its anastomosis with the right pulmonary artery. The proximal subclavian artery was large but not aneurysmal, and the anastomotic site itself was of normal caliber. The shunt flow was large, producing almost normal peripheral oxygen saturation (93%) and moderate pulmonary artery hypertension (pulmonary artery pressure 50/30 torr).

At operation on October 13, 1980, a 5 to 6 cm aneurysm of the distal subclavian artery was found. After the institution of cardiopulmonary bypass, the aneurysm was dissected free and opened. The subclavian artery proximally and the pulmonary artery distally were over-sewn and the aneurysmal sac was excised. Correction of the intracardiac defects was then performed. The postoperative recovery was uneventful with the exception of right vocal cord paralysis which has persisted. A 1 year postoperative chest X-ray film revealed the resolution of the mediastinal mass, and the patient remains asymptomatic almost 3 years after repair (Fig. 1B).

Case 2. A 48 year old woman with a diagnosis of tetralogy of Fallot had a right Blalock-Taussig shunt placed

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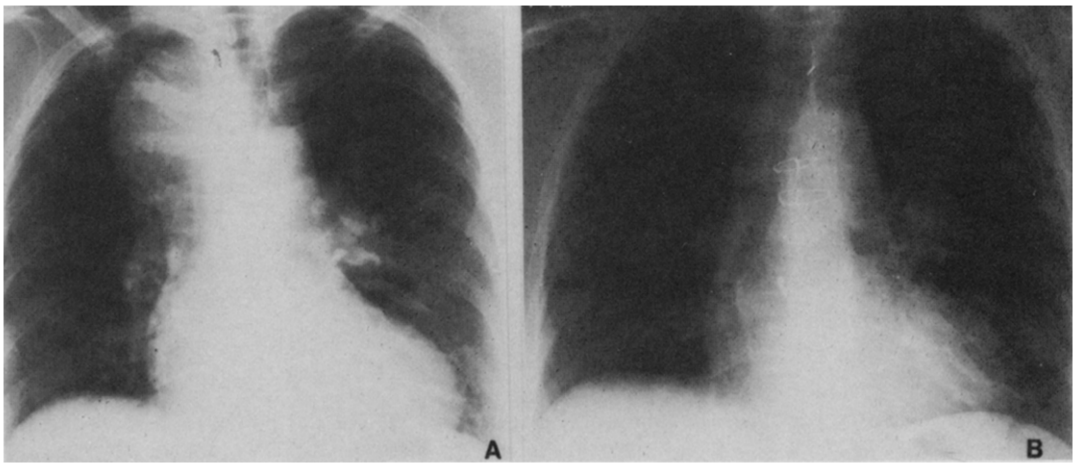


Figure 1. Case 1. Chest roentgenograms before **(A)** and 3 years after **(B)** repair of the aneurysm. Note decrease in width of superior mediastinum.

for cyanosis during 1950 when she was 14 years old. She subsequently did well until the age of 47 years, when she developed progressive symptoms of congestive heart failure. Cardiac catheterization revealed very mild peripheral artery desaturation (86%), moderately severe pulmonary artery hypertension (pulmonary artery pressure 70/40 torr) and the classic findings of tetralogy of Fallot (Fig. 2A). Arteriography demonstrated an aneurysmal Blalock-Taussig shunt. She underwent complete correction of her cyanotic congenital heart disease and an unsuccessful attempt at ligation of the shunt. Reconstruction of the right ventricular outflow tract required a valved conduit (Hancock Laboratories), because of the relation of the large ventricular septal defect to the aorta and pulmonary artery. Her postoperative recovery was slow, complicated by congestive heart failure and arrhythmias.

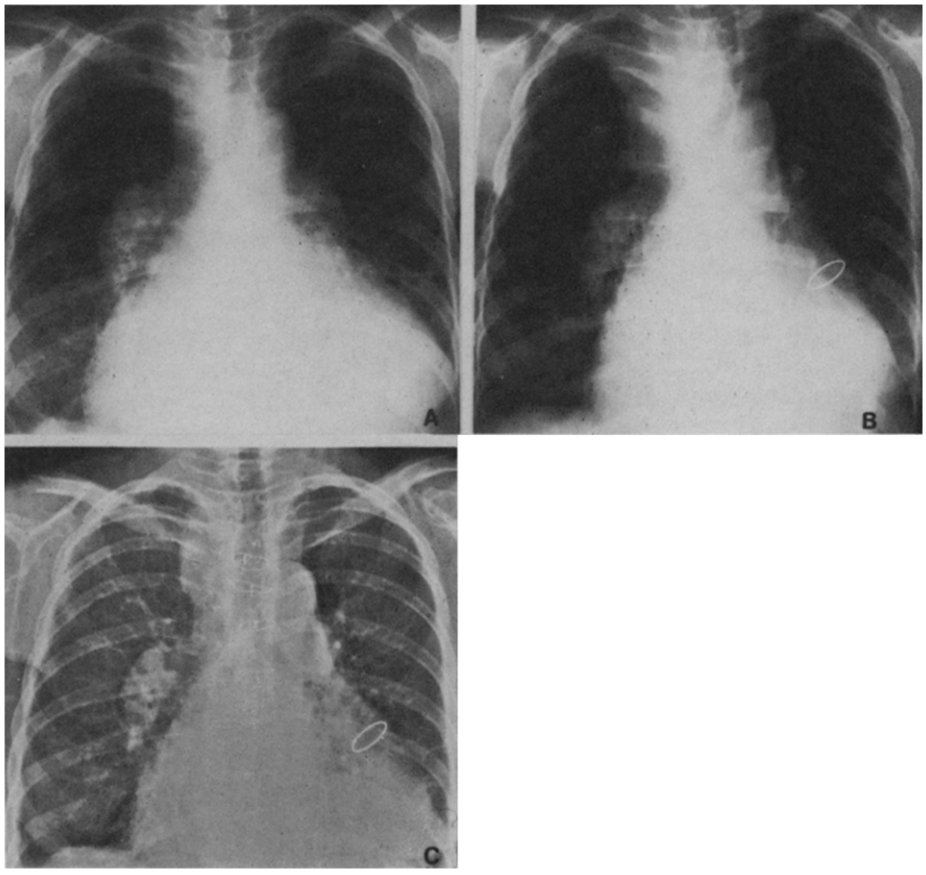


Figure 2. Case 2. Chest roentgenograms before **(A)** and after **(B)** correction of tetralogy of Fallot, but before repair of aneurysm. **C,** 5 months after repair of aneurysm. Note the partial resolution of superior mediastinal widening.

Nine months after total operative correction, the patient returned with a 2 month history of intermittent throbbing pain at the base of the right side of the neck associated with the development of edema of both eyelids and mild hoarseness. No neurologic symptoms were noted, nor did she complain of congestive symptoms, headache, dysphagia or dyspnea.

On physical examination, a 3 to 4 cm pulsatile mass extending from the thorax was palpated at the base of the right side of the neck under the medial head of the sternocleidomastoid muscle, deviating the trachea to the left and extending under the manubrium and clavicle. There was mild edema of both upper eyelids, and right vocal cord paralysis was confirmed by indirect laryngoscopy. A chest X-ray film revealed a widened superior mediastinum without much definable change (Fig. 2B). Aortography revealed a patent Blalock-Taussig shunt with aneurysmal dilation, but the subclavian artery was not well defined without selective injections.

The patient had a progression of symptoms and an increase in size of the aneurysm by computed tomography over a 2 month period. Selective angiography (Fig. 3A) confirmed the increase in size of the aneurysm, measuring 5 × 12 cm, with calcification of the wall. The entire shunt was aneurysmal, with the exception of a 1 to 2 cm length of subclavian artery at the origin from the innominate artery and the pulmonary artery anastomosis.

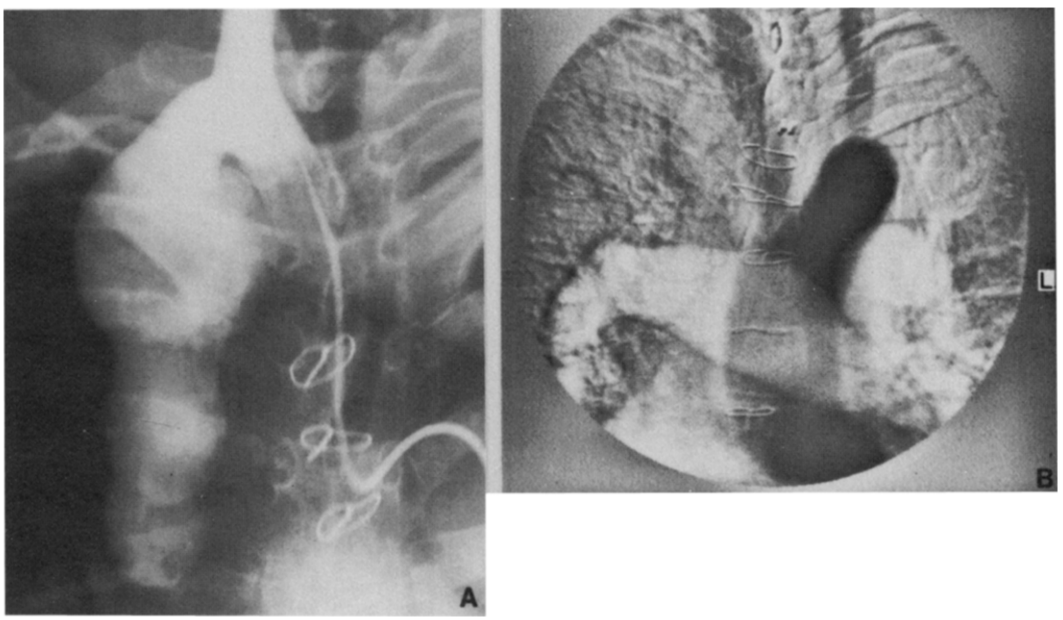
Figure 3. Case 2. **A,** Selective arteriogram of innominate artery before repair of the Blalock-Taussig shunt aneurysm. Note the large aneurysm and uninvolved proximal segment of subclavian artery. **B,** Digital subtraction angiogram 5 months after repair of the aneurysm with division of the proximal subclavian artery. Note normal pulmonary artery anatomy.

The patient was explored through the previous median sternotomy incision, which was extended up into the right side of the neck. The innominate vein was divided, and the innominate artery identified and traced to the bifurcation. Just beyond the origin of the right subclavian artery, a large aneurysm was palpated, extending medially toward the aortic arch and almost occluding the superior vena cava. The full extent of the aneurysm could not be assessed as it extended behind the arch and down into the mediastinum. The subclavian artery, between its origin and the aneurysm, was divided between vascular clamps and oversewn. Her recovery was uneventful.

Five months postoperatively the patient was asymptomatic. There was no pulsatile mass in the right neck, no facial edema and the right vocal cord moved normally. Chest X-ray film revealed a decrease in superior mediastinal width (Fig. 2C). Digital subtraction angiography revealed a normal right pulmonary artery and no opacification of the aneurysm (Fig. 3B).

Discussion

Since 1945 when this shunt was initially described (11), many subclavian to pulmonary artery shunts have been created. Although substantial progress has been made in the operative correction of cyanotic congenital heart disease since that time, a number of these patients are still living with a patent Blalock-Taussig shunt (4,19); in addition, there are still substantial numbers of patients undergoing creation of such a shunt for a variety of reasons (2–4,15–21). Some of these patients have complex lesions that are not easily corrected and will live with their shunt for a long



period of time. There have been a number of early and late complications of Blalock-Taussig shunts reported (2–4,18–21).

Cause of aneurysm. Subclavian artery aneurysms are rare in situ (22,23). There has only been one previously reported case (17) of a subclavian artery aneurysm associated with a subclavian to pulmonary artery anastomosis, although there have been reports (2,24,25) of aneurysmal dilation of the pulmonary artery after creation of a Blalock-Taussig shunt. Our report details the operative correction of that first case and the recent addition of a second case. Several historical points are common to both patients. Both presented more than 30 years after creation of their shunt and both had moderate pulmonary artery hypertension with high peripheral artery oxygen saturation suggestive of a large shunt flow. This indicates that duration of the shunt and magnitude of shunt flow may play a role in the development of this rare late complication. This relation has been suggested to explain the late development of pulmonary artery hypertension after the creation of a Blalock-Taussig shunt as well (2,20). Operative trauma to the subclavian artery has been suggested as the cause of the subclavian artery aneurysm (17), but although possible, this seems less likely because of the rarity of this lesion and the extremely late appearance of the aneurysm (more than 30 years).

Management of aneurysm. As with peripheral artery aneurysms in other sites, a subclavian artery aneurysm may occlude, embolize or rupture (26). Rupture is rare but does occur (26). Elective surgical repair is currently believed to be the most prudent treatment (23). In our second patient, the large, symptomatic and expanding aneurysm was clearly documented, and there is little controversy concerning operative intervention. In Case 1, the asymptomatic aneurysm was followed up with serial chest X-ray films for 4 years without an appreciable increase in size before elective repair was carried out. These aneurysms, however, may enlarge appreciably without increasing the mediastinal silhouette; therefore, it may not be clear whether there is any progression in size in the aneurysm over time. Computed tomography should be superior for assessing change in aneurysm size.

If symptoms are present or if the aneurysm is large or expanding, the decision to intervene is straightforward, either combined with total correction of the intracardiac defects or with creation of another shunt. In the patient with a small to moderate-sized aneurysm, the decision to operate is more difficult. If total operative correction of the intracardiac defects can be accomplished, then the presence of the aneurysm would prompt early total correction and aneurysmectomy. If, however, total correction is not feasible, the patient should be followed up with serial computed tomographic scans; the development of symptoms related to the Blalock-Taussig shunt aneurysm or enlargement of the aneurysm could then be used as a criterion for operative intervention.

Surgical approach. If adequate pulmonary flow is established, either by intracardiac correction or another shunt, the simplest operative approach is division and oversewing of the proximal subclavian artery through a median sternotomy. This approach reduces the likelihood of recurrent laryngeal nerve injury and might also be advantageous at the time of concomitant intracardiac repair if difficulties should develop in proximal or distal control of the aneurysm. This was the technique of repair in Case 2, the rationale being that subsequent thrombosis of the aneurysm will result in fibrous obliteration and contraction of the space-occupying mass. Such was the case in our patient, and this corresponded with complete resolution of all symptoms and return of recurrent laryngeal nerve function. Pulmonary artery flow was not compromised, and no evidence of pulmonary emboli was noted (Fig. 3C).

Implications. The possibility of a Blalock-Taussig shunt aneurysm should be considered when a patient who has had a shunt for a long period of time presents with signs or symptoms of compression of mediastinal structures or an enlarging superior mediastinal mass. Confirmation of the diagnosis can be made by computed tomography or angiography, and a decision can then be made with respect to when to proceed with surgical intervention. A simple and safe approach to correct this lesion is division and oversewing of the proximal subclavian artery through an anterior approach, assuming adequate pulmonary blood flow is already present or can be established concomitantly.

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