

THE SURGICAL TREATMENT OF SUPRAVALVULAR AORTIC STENOSIS BASED ON TWO CASES

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(RECEIVED FOR PUBLICATION JANUARY 17, 1962)

An increasing number of cases of supra-avalvular aortic stenosis have been reported in recent years. In those hitherto described, the stricture has consisted of either a fibrous membrane directly adjacent to the cusps or a true stenosis at the level of the upper border of the sinus of Valsalva where, as mentioned by Kreel, Reiss, Strauss, Blumenthal, and Baronofsky (1959), a slight, circular inward bulge in the aortic wall is normally present. Formerly, attempts at operative treatment of supra-avalvular aortic stenosis met with failure. However, in 1961, McGoon, Mankin, Vlad, and Kirklin reported three cases in which operation had been performed at the Mayo Clinic with excellent results. In these the supra-avalvular stenosis had been split and a patch graft sutured into the defect. Starr, Dotter, and Griswold (1961) also reported a patient successfully operated on by the same technique. During the past two years two patients who had supra-avalvular aortic stenosis have successfully undergone operation at the Thoracic Clinic, Karolinska Sjukhuset; an account of them seems to be warranted.

CASE REPORTS

CASE 1.—A 28-year-old woman had had organic heart disease diagnosed in childhood. Her physical activity was slightly limited. A high-frequency, early to mid-systolic murmur was audible, with a maximum over the second right interspace. The heart volume was 430 ml./m.² of body surface. The E.C.G. showed left ventricular hypertrophy. At cardiac catheterization the pressure was 233/19 mm. Hg in the left ventricle and 111/61 mm. Hg in the aorta (i.e., indicative of tight aortic stenosis). In addition, there were signs of moderate pulmonary hypertension with raised pulmonary resistance. No angiocardigraphic examination was made and the condition was diagnosed as congenital aortic stenosis.

Operation was performed on September 16, 1958, under extracorporeal circulation with the heart-lung machine of Crafoord and Senning. A few centimetres above the aortic valve the wall of the ascending aorta was abnormal; the vessel was partly occluded

by a thickening, of which the main part protruded into the lumen of the aorta. As much as possible of this obstruction was removed, making an opening in the lumen of the aorta slightly wider than an index finger. The aortic cusps were normal. The aortic wall was then sutured. Post-operatively, a systolic murmur of the same type and localization as was heard before operation was present, but it was fainter, of lower frequency, and less harsh. The patient was discharged from hospital in good condition just over three weeks after operation.

She was readmitted to hospital for follow-up examination in September, 1961. She had been asymptomatic since operation, and worked as an assistant hospital nurse on night duty. In 1960 she had given birth to a child. Both pregnancy and delivery were uncomplicated. The heart volume was 400 ml./m.² of body surface compared with 430 ml./m.² pre-operatively. The E.C.G. no longer showed hypertrophy of the left ventricle, and depression of the S-T intervals and T waves was less. At phonocardiography a medium-frequency, diamond-shaped pansystolic murmur was recorded, with a maximum over the second right interspace, but of lower intensity than previously. Angiocardiology showed that a stricture with an inner diameter of 1.5 cm. was still present at the site of the supra-avalvular stenosis (Fig. 1). The systolic pressure gradient across the stenosis was, however, only 37 mm. Hg compared with 122 mm. Hg pre-operatively (Table I). Consequently, we felt that there was no indication for further intervention.

TABLE I
PRE- AND POST-OPERATIVE CARDIAC CATHETERIZATION IN TWO CASES OF SUPRAVALVULAR AORTIC STENOSIS

	Pressure (mm. Hg)					
	Pre-operative			Post-operative		
	Left Ven-tricle	Aorta	Systolic Gradient	Left Ven-tricle	Aorta	Systolic Gradient
Case 1*	233 19	111 61	122	141 0-6	104 52	37
„ 2*	262 3-5-24	128 79	134	132 8-29	108 68	24

* In Case 1, operation consisted of local excision of the stenosis; and in Case 2, of splitting the aorta through the stenosis and insertion of a patch graft.

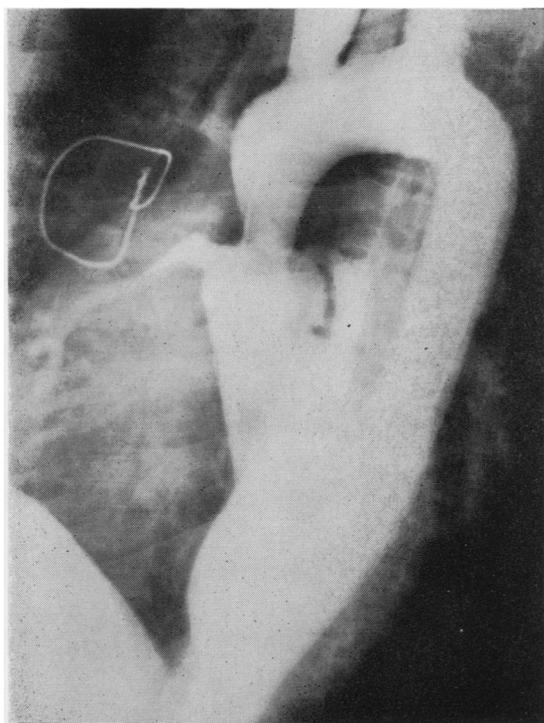


FIG. 1a

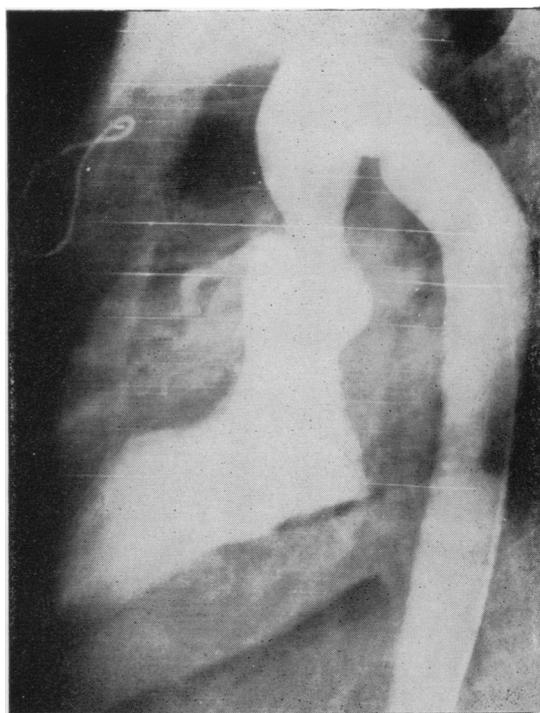


FIG. 1b

FIG. 1 (a) and (b).—Case 1. Post-operative angiocardiology (Dr. Eric Carlsson, Thoracic Clinic) in two planes in a case of supra-valvular aortic stenosis. Operation consisted of local excision of the stenosis. Post-operative pressure gradient across the former site of stenosis 37 mm. Hg.

CASE 2.—An 8-year-old boy had congenital organic heart disease but no appreciable functional limitation. At the age of 7 he had had a subarachnoid haemorrhage. Investigation at another hospital in 1959 disclosed a systolic thrill over the whole praecordium transmitted to the vessels of the neck, as well as a high-frequency systolic murmur (grade 6) with a maximum over the second right interspace. Although the peripheral pulsations were faint, the blood pressure could be measured in both the arms and legs (100/75 mm. Hg). The heart volume was 620 ml./m.² of body surface. Moderate stasis was present in the pulmonary circulation. At cardiac catheterization the pressure was 262/3.5–24 mm. Hg in the left ventricle and 128/79 mm. Hg in the aorta. Angiocardiology showed marked hypertrophy of the left ventricle as well as stenosis of the ascending aorta for slightly over 1.5 cm., starting a few centimetres above the plane of the aortic valve. At the narrowest site the lumen was 6 mm. in diameter (Fig. 2). Among the findings at cerebral angiography was an aneurysm of the posterior cerebral artery. It was decided to operate on the aortic stenosis in the first place, this being the more vital indication.

Operation was performed on June 14, 1959, under extracorporeal circulation with the heart-lung

machine of Crafoord and Senning. The ascending aorta was found to be greatly stenosed a few centimetres above the valvular plane. An anterior incision was made in the vessel, extending from above the coarctation to below it. At the narrowest site the diameter of the lumen was 6 mm. The aortic cusps were normal. A diamond-shaped dacron graft, 3.5 cm. long and 2.5 cm. wide, was sutured into the incision in the aorta. After an uneventful post-operative course the patient was sent on the nineteenth day to another hospital for further investigation of the cerebral vascular anomalies.

He was readmitted for follow-up examination in August, 1961, two years after operation. No operation had been performed for the cerebral vascular anomalies since it was considered to be too risky. The boy's physical activity was normal. His heart volume was 400 ml./m.² of body surface as compared to 620 ml./m.² pre-operatively. A faint (grade 3) systolic murmur was audible with a maximum over the second right interspace. Angiocardiology, with injection of contrast medium into the left ventricle, showed that there was no longer any impediment to passage in the ascending aorta. An indentation about 3 mm. deep was visible, corresponding to the site of the graft (Fig. 3). The pressure conditions at cardiac catheterization are seen in Table I.

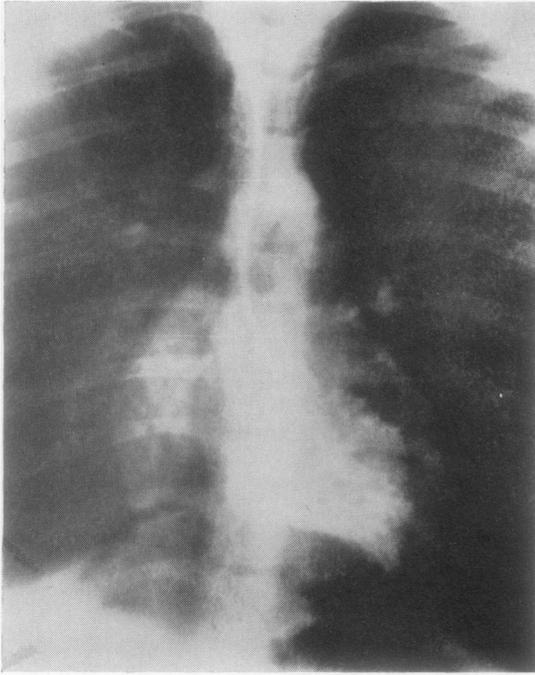


FIG. 2a

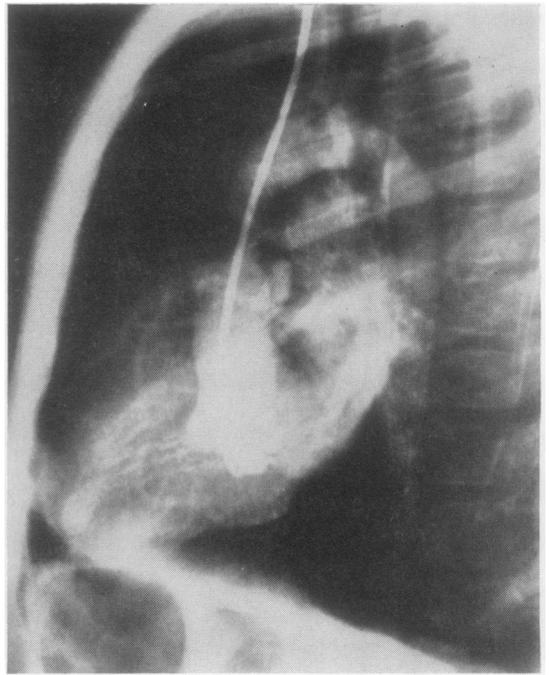


FIG. 2b

FIG. 2 (a) and (b).—Case 2. Pre-operative angiocardiography in two planes in a case of supravalvular aortic stenosis. Pre-operative pressure gradient across the stenosis 134 mm. Hg. (Courtesy of Dr. G. Jönsson, Södersjukhuset, Stockholm.)

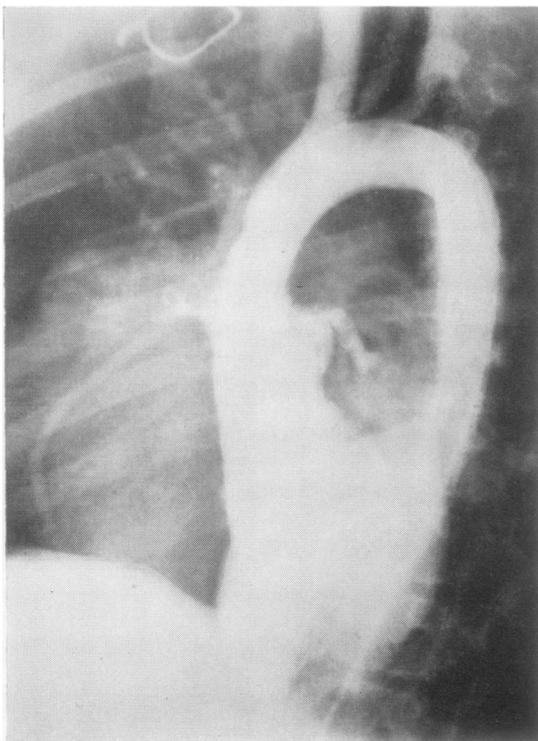


FIG. 3a

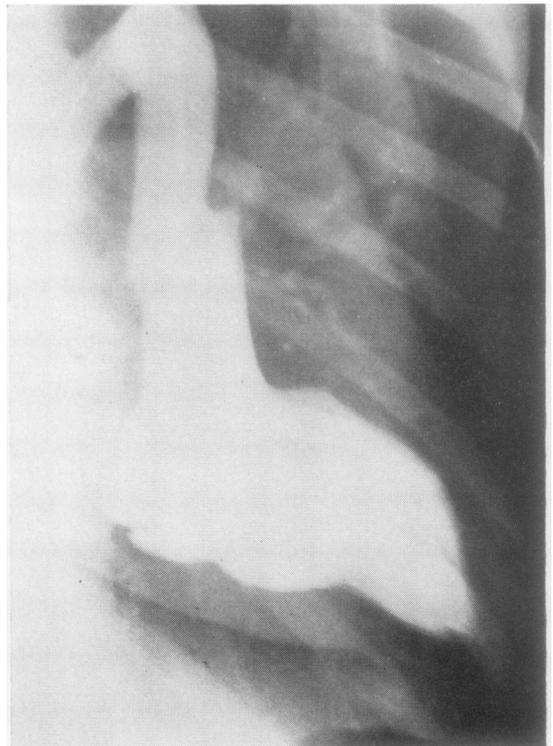


FIG. 3b

FIG. 3 (a) and (b).—Case 2. Post-operative angiocardiography (Dr. Eric Carlsson, Thoracic Clinic). Operation consisted of splitting the aorta through the stenosis and insertion of a patch graft. Post-operative pressure gradient 24 mm. Hg.

DISCUSSION

In both our cases the stenosis was at the typical site, i.e., at the upper border of the sinus of Valsalva. A remarkable feature in Case 2 is that the stricture involved a segment of the aorta more than 1.5 cm. long. Both radiologically and macroscopically it resembled a coarctation of the aorta. No biopsy was done. In Case 1 the stenosis was removed and examined microscopically. It was found to contain smooth muscle, indicating that inward bulging of the media was responsible for the stenosis. According to Edwards (1953) this medial deformity is the anatomical basis of constriction of the lumen in true coarctation of the aorta.

In 1959 Eiken described a case of coarctation of the aorta central to the origin of the innominate artery. He stated that he had been unable to trace any case described earlier in the literature. Eiken's patient was a 35-year-old woman with such pronounced calcification of the aortic wall that the coarctation was directly visible on plain radiographs and was easily depicted by tomography. In view of the advanced calcifications her condition was regarded as inoperable. Consequently no microscopical diagnosis was available.

At physical examination these cases present the same features as cases of valvular aortic stenosis. The diagnosis is established by the withdrawal curve on catheterization from either the aortic side or the left side of the heart. The most certain way of obtaining a diagnosis is by angiocardiology or aortography. In our Case 1 pre-operative angiography had not been performed. The condition was diagnosed as tight aortic stenosis, on the basis of the physical findings and the results of catheterization. At operation we were surprised to find completely normal aortic cusps and an aorta of ordinary width in the valvular plane. We found instead a stenosis of the diaphragm type with thickening of the aortic wall above the valve. In Case 2 the anatomical conditions had been depicted at pre-operative angiocardiology, and we could therefore make a direct attack on the malformation at operation. In the case reported by Kreel *et al.* there was not only supra-valvular aortic stenosis but also valvular lesions the nature of which could not be identified at operation and which led to the patient's death.

With the technique now used for angiocardiology, this examination is associated with a relatively small risk for the patient. Consequently, in our opinion, all patients with physical signs of

valvular aortic stenosis should undergo an angiocardiology examination. This is partly to avoid unexpected coincident anomalies and partly, but not least, to obtain an exact pre-operative diagnosis. We, in contrast to McGoon *et al.*, do not believe that, because planning of the operation is the same in the presence of subvalvular, valvular, and supra-valvular aortic stenosis, this is sufficient reason to abstain from pre-operative angiocardiology.

SURGICAL TECHNIQUE.—The operation necessitates use of the heart-lung machine. The most suitable surgical procedure seems to us to be that applied in our Case 2, which is essentially the same as that described by Starr *et al.* and by McGoon *et al.* After a midline incision and cleaving of the sternum, the venae cavae and one of the femoral arteries are cannulated. Continuous deloading of the left side of the heart is procured by a cannula in the left atrium. When perfusion has been started, the aorta is occluded and an arched incision is made in its anterior wall through the stricture and well down into the non-coronary aortic sinus. A diamond-shaped dacron or teflon graft of appropriate size is then sutured into the defect. The technique is analogous to that described by Senning (1959), among others. The graft should be large enough to cover the whole incision, with its widest part at the level of the most stenosed segment. An adventitial flap, prepared in advance, can be swept over the graft and suture lines.

It is important for the suction cannula in the left atrium to be shut off before suturing of the graft is finished. This gives the left side of the heart time to become filled with blood, with expulsion of all air that may have been sucked into the left atrium. Use of the left atrial cannula provides an operative field that is as bloodless as possible. Consequently, the ordinary coronary sucker tip, which masks the operative field, need be used only to a limited extent, as Starr *et al.* have also pointed out. Separate coronary perfusion seems to be superfluous as occlusion scarcely requires to exceed 15 to 20 minutes.

The surgical technique used in our Case 1 does not appear to have been described earlier. When the follow-up examination was made, we were prepared to reoperate on the patient, with insertion of a graft. Since she was, however, asymptomatic and had a decreased heart volume and a systolic pressure gradient of only 37 mm. Hg, we regarded the results as highly satisfactory.

SUMMARY

An account is given of two patients with supra-valvular aortic stenosis, in whom operation was successfully performed by different methods. The surgical technique is discussed, and widening of the stenosed supra-valvular segment with a patch graft is recommended. The importance is stressed of exact pre-operative visualization of the anatomical conditions by angiocardiography. The excellent results of operation could be verified in

both cases by follow-up examination three and two years later, respectively.

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