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Neoaortic Aneurysm After Stage I Norwood Reconstruction

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Treatment of hypoplastic left heart syndrome through staged repairs has resulted in patients surviving into adulthood. Use of either aortic or pulmonary homografts in performing the neo-aortic reconstruction has become the standard of practice with relatively few problems. We report the case of an asymptomatic adolescent boy who had an enlarging neo-aortic aneurysm and mild neo-aortic regurgitation develop after undergoing a stage I Norwood procedure using a pulmonary homograft. Given the risk for rupture and a concern for further functional deterioration of the neo-aortic valve, the patient underwent repair. Histologic examination showed a striking accumulation of myxoid material as well as abnormal vasculature in both the native and engrafted portions of the neo-aorta.

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Establishment of adequate systemic perfusion, as part of the first stage in the treatment of hypoplastic left heart syndrome, relies on the transformation of the pulmonary valve into a systemic valve along with aortic reconstruction using either an aortic or pulmonary homograft. The effect of exposing the native pulmonary valve apparatus to systemic pressure has generated significant discussion as to its long-term functional durability and potential dysfunction. Dilation of the sinotubular junction has been suggested as one mechanism for this dysfunction. Aneurysmal dilatation of homograft tissue in this setting has not been reported previously, however, experimental data suggests that pulmonary homografts possess a susceptibility for dilatation not seen in their aortic counterpart [1, 2]. Therefore, the choice of homograft can potentially impact the functional durability of the neo-aortic valve.

The patient is a 14-year-old boy who he underwent a stage I Norwood procedure using a pulmonary homograft at 3 days of life. At 17 months of age he completed his staged repair by proceeding directly to a lateral tunnel fenestrated Fontan procedure. At 9 years of age, monitoring echocardiography revealed dilatation of his neo-aorta to a diameter of 4.2 cm in greatest dimension. During the following 5 years, the neo-aorta progressively enlarged, but the patient remained asymptomatic. In addition, the patient's neo-aortic valve was noted to be mildly regurgitant despite no enlargement in the neo-aortic valve annulus. Magnetic resonance imaging demonstrated the neo-aorta to be 5.5 cm in

the antero-posterior dimension and 7.0 cm in the left-to-right dimension (Fig 1).

Given this dilatation, along with the dysfunction of the neo-aortic valve, the patient underwent replacement of the neo-aortic aneurysm with a 26-mm Hemashield woven Dacron conduit (Boston Scientific Corp, Natick, MA). The distal anastomosis was performed under hypothermic circulatory arrest. Intraoperatively the neo-aortic annulus was within normal limits. By using a 26-mm conduit, we effectively downsized the sinotubular junction of the neo-aorta with the intention of improving or at least preventing further functional deterioration of the neo-aortic valve.

Gross examination of the pathologic specimen revealed fragments of the vessel wall ranging from 0.2 to 0.4 cm in thickness. The endothelial surface was tan, glistening, and largely smooth with foci of irregular tan plaques and pits. Mural calcification was focally palpable and the suture material was embedded in the wall. Distinction between the native vessel and the homograft was not grossly possible.

Microscopic examination of hematoxylin-eosin and elastic tissue stains revealed areas interpreted as native aorta and areas interpreted as pulmonary homograft. The aorta showed numerous wavy parallel elastic laminae throughout the media and extending into the intima. In most areas, the aorta showed cystic medial degeneration and fibrointimal proliferation. Adventitial arteries showed prominent medial hypertrophy. The pulmonary homograft showed only a thin rim of heavily fragmented elastic fibers, located immediately adjacent to the adventitia. Both the adventitia and this elastic tissue layer showed dense fibrous scarring and numerous ectatic thin-walled blood vessels (Fig 2A). The remainder of the vessel wall consisted of paucicellular myofibroblastic cells uniformly distributed in a fibrous matrix that was similar to the native aortic wall in thickness and also showed abundant myxoid material in areas (Fig 2B). The presence of suture granulomas helped to delineate the transition between native tissue and the graft.

The patient's postoperative course was unremarkable. He was discharged home on postoperative day 14. He has returned to his normal level of preoperative activity; however, his neo-aortic regurgitation failed to resolve.

Comment

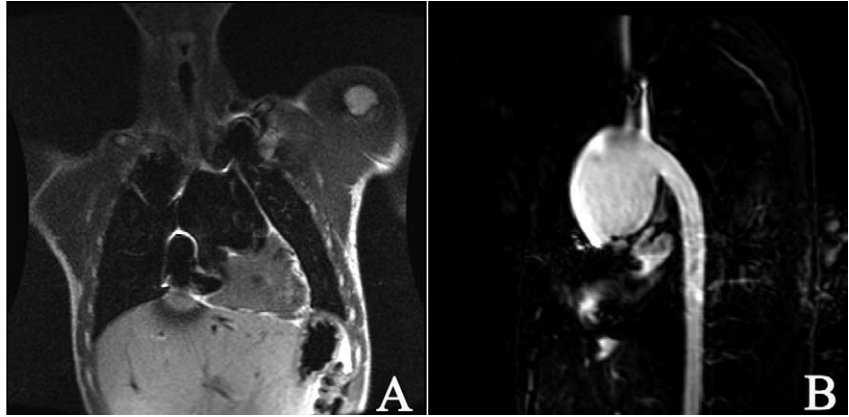
Aortic reconstruction in patients with hypoplastic left heart syndrome generally requires patch augmentation of the aorta along with incorporation of the pulmonary valve into the systemic circulation. The development of neo-aortic regurgitation and neo-aortic aneurysm in patients after stage I may be related to the patch material used, the pulmonary valve, and its sinotubular component, or a combination of these factors. Both rendered the neo-aorta and its valve susceptible to the changes observed in this case, which was corroborated by both clinical and experimental data.

Despite the application of the stage I Norwood procedure by many institutions, there have been no reports documenting aneurysmal dilatation after homograft patch augmentation of the aorta. Little has been reported about the long-term durability of the patch material used in this setting; however, experimental data has demon-

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Fig 1. Magnetic resonance imaging. (A) Anteroposterior view of native and neoartical roots along with Stansel connection and aneurysm. (B) Saggital view.



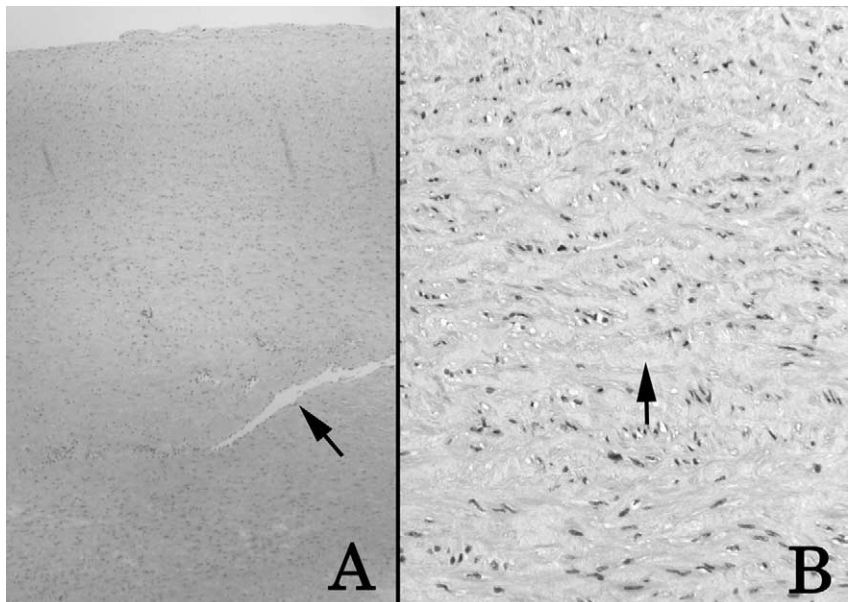
strated a higher incidence of aneurysm formation for pulmonary homografts [1, 2].

The pulmonary homograft included a clearly viable layer of myofibroblastic cells resembling a nonelastic vascular media intima. The accumulation of myxoid extracellular matrix in this layer was striking. The native aorta also showed myxoid degeneration as a major histologic feature. This material has been noted as a non-specific finding in aortic aneurysms due to aging, arteriosclerosis, and Marfan's syndrome, and in animal homograft models, but we believe it has not been reported in a human pulmonary homograft [3]. We noted that the vascular supplies in both the native aorta and graft were abnormal. The aortic arch is typically well vascularized and rich in vasa vasora. The native aortic portion of this patient's neoaorta showed marked medial hypertrophy of adventitial vessels, perhaps a consequence of longstanding surgical disruption or perhaps due to the underlying congenital abnormality. The homograft, which attained a medial intimal thickness comparable with that seen in the aorta, was served by

thin-walled ectatic vessels that appeared to have arisen out of granulation tissue in the adventitia and deep media. It is interesting to speculate whether a compromised or outstripped vascular supply may have been responsible for much of the degenerative change seen in both components of the neoaorta.

Our case contrasts with the report by Mahle and colleagues, which suggested appropriate growth of the aortic arch after Norwood reconstruction with homograft tissue. Mahle and colleagues [4] found that growth occurred in the native aorta. Our case also clearly documented growth of the native aorta though the histologic findings, which suggests that at least in part, aneurysmal dilation was secondary to degeneration within the aorta. Although the risk of rupture or dissection is unknown, we believe that it was reasonable to recommend surgical intervention when the dimensions had reached 5.5 and 7.0 cm rather than a Z score indication for surgery because of lack of normative values in this setting. Nevertheless it was felt that these dimensions more than likely represented a Z score that would be equivalent to

Fig 2. Pulmonary homograft showing (A) a thick intima media composed of myofibroblastic cells and containing thin-walled ectatic blood vessels (arrow) ($\times 40$), and (B) accumulation of pale myxoid extracellular material (arrow) between the cells ($\times 200$).



at least +10, which is the level at which we recommend intervention for aortic root aneurysms for Marfan's syndrome.

Exposure of the pulmonary valve to systemic pressure is also seen in the Ross procedure and arterial switch operation. Where as no data exists as to the long-term durability of the pulmonary valve in the Norwood procedure, data from the Ross and arterial switch operation have demonstrated a potential for valvular regurgitation that is associated with dilation of the sinotubular junction [5, 6]. Therefore the finding of mild neo-aortic valve regurgitation in our patient can likely be explained by the dilation of the sinotubular junction due either to the native deterioration of this region or the dilation of homograft material immediately distal to it. We speculated that the progressive nature of this process, in association with the aneurysmal component of the neo-aorta, created the likelihood that the dysfunction of the valve would further progress as the aneurysm grew larger. As a result, by intervening, we aimed not only to prevent potential aneurysmal rupture, but also to correct or at least prevent further deterioration of valve function.

Given these observations, it is difficult to discern whether dilation of the neo-aorta was due to the material used or a more general response to an altered physiologic state in the setting of a reconstructed ascending aorta and arch aorta. Histologically, there were no particular similarities between what was seen in the pulmonary homograft of the patient and the observations made in the experimental pulmonary homograft aneurysms. Either way, it would be difficult to make any conclusions

given the scope of this comparison. If aneurysm formation were a more general response to the altered physiologic state in the setting of reconstructed anatomy, one would expect a greater frequency of this phenomenon. The finding of cystic medial degeneration in the native aorta suggests that the culprit may be a more generalized response rather than a tissue-specific effect. If this were to hold true, we can expect reports of similar cases in the coming years, as this patient population continues to age.

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