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Original Article

Can fibrotic bands in the aortic arch cause innocent murmurs in childhood?

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Abstract Children with innocent murmurs are often referred to a paediatric cardiologist for diagnosis. The most common murmurs of early childhood are the so-called Still's murmurs, followed by ejection murmurs across the pulmonary or aortic vessels, and the venous hum. There also exists a high coincidence of murmurs with the presence of tendinous structures traversing the cavity of the left ventricle. In this report, we describe 6 patients who presented to our outpatient clinic with cardiac murmurs. None of them had abnormalities on the clinical examination, electrocardiographic, or echocardiographic investigation. They presented a similar murmur that was also audible over the back. On closer examination of the aorta with cross-sectional echocardiography, we discovered echogenic, tendinous structures crossing the lumen of the descending aorta or the aortic arch. Whilst we are not yet able to prove that the cords produced the innocent murmurs, the association is highly suggestive.

Keywords: Innocent murmurs; paediatric cardiologist; examination; technical skills; non-invasive methods

INNOCENT MURMURS IN CHILDHOOD ARE TYPICALLY associated with neither anatomical nor physiological anomalies. Thus, in most patients with such murmurs, which are early-systolic, soft, and heard only over a small area, there are no clinical symptoms nor diagnostic signs. Such murmurs may be the so-called Still's murmur, producing an early systolic, soft and vibratory sound at the left sternal border, or ejection murmurs across the pulmonary or aortic vessels.[1] There also exists a coincidence between innocent murmurs and tendinous structures crossing the cavity of the left ventricle. These cords are typically singular, and run from the lateral wall of the left ventricle to the septum. Such echogenic threads are reported in seven-tenths of echocardiograms performed on children with innocent murmurs.[2] The resulting murmur is of middle frequency with a musical sound, and is heard best in the 3rd and 4th intercostal space at the left sternal border.

Thus far, such tendinous structures have been described in the aortic arch only subsequent to post-mortem examinations (Fig. 1). Over a period of 6 months, we have seen 6 patients referred to our clinic for evaluation of innocent murmurs in whom such tendinous structures were discovered in the aortic arch.

Patients studied

The 6 patients, aged from 5 to 22 years, presented over a period of 6 months to our out-patient clinic in the Department of Paediatric Cardiology of the University Children's Hospital, Göttingen, for workup of a murmur of unknown origin. The patients were healthy and active, without other abnormalities on clinical examination. In all patients, a vibratory early systolic murmur was heard, loudest in the second intercostal space on the left sternal border, with radiation to the lower neck. The murmur was also heard under the scapula over the left side of the back. No pathological findings were found on the

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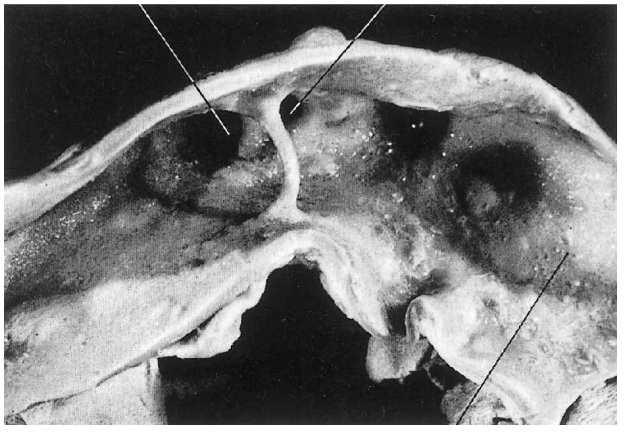

Figure 1.
Anatomical picture of the aortic string reaching across the lumen of the aorta (with kind permission of Verlag Urban und Vogel).

Figure 2.
Suprasternal view with colour Doppler showing the separation of the Doppler flow by the "string", AAo: ascending aorta; TR: brachiocephalic trunk; string: showing the fibrous band in the colour-Doppler signal; DAo: descending aorta.

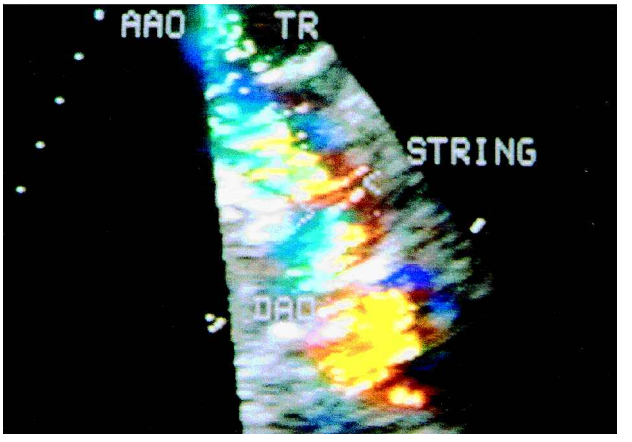

Figure 3.
Colour-Doppler interrogation reveals the separation of streams in the aortic lumen.

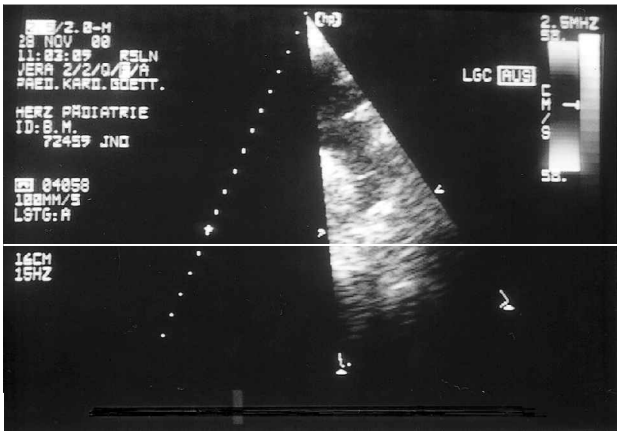

Figure 4.
Suprasternal echocardiography in the sagittal view of the aortic arch reveals a "string" located between the left common carotid artery and the subclavian artery.

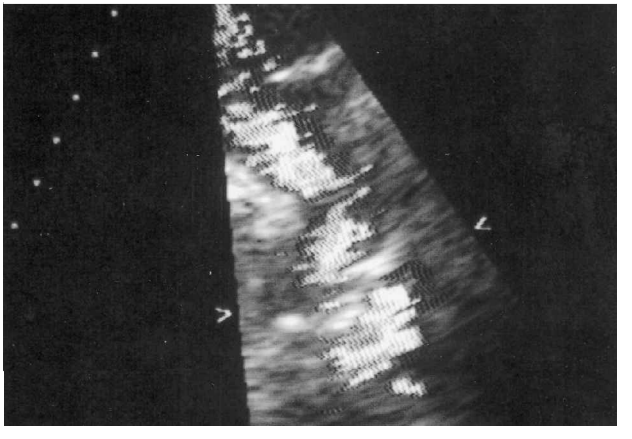

Figure 5.
Enlargement of Figure 4 showing the separated flows as revealed by Doppler interrogation.

electrocardiogram. Congenital cardiac malformations, and tendinous structures in the left ventricle, were excluded echocardiographically. Normal velocities of flow were recorded across both the aortic and the pulmonary valves. Initial images of the ascending aorta and the aortic arch also appeared normal on cross-sectional examination. Subsequent interrogation using colour echocardiography, however, revealed turbulent flow in the descending aorta (Figs 2 and 3). Closer examination of the cross-sectional images then demonstrated echogenic, tendinous structures, crossing either the lumen of the aortic arch or the descending aorta (Fig. 4). The previously laminar profile of aortic flow was interrupted and became turbulent at the location of these structures (Fig. 5). In two children, the strings seemed like 'wavebreakers' splitting the flow of blood (Fig. 6). These cords were found at

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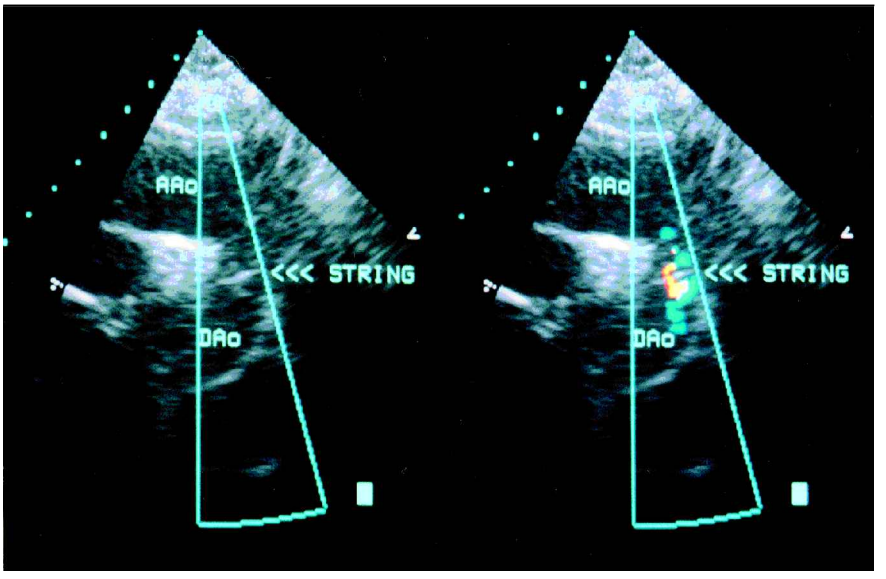

Figure 6.
Cross-sectional imaging of a string in the aortic arch, with flow of blood separated by the string. AAo: ascending aorta; DAo: descending aorta.

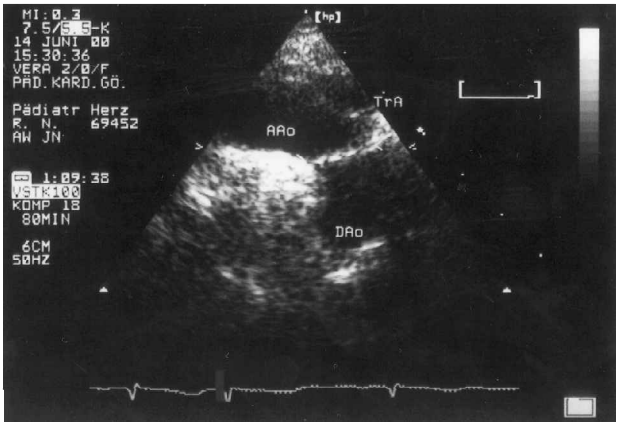

Figure 7.
Cross-sectional evidence of a string in the aortic arch. AAo: ascending aorta; TrA: brachiocephalic trunk; DAo: descending thoracic aorta.

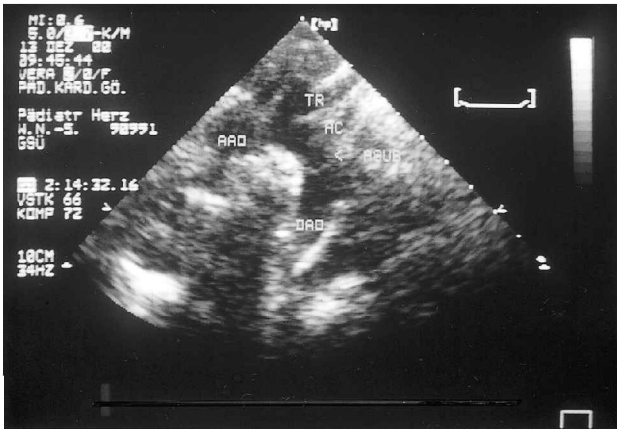

Figure 8.
Echocardiographic findings from the suprasternal notch. AAo: ascending aorta; TR: brachiocephalic trunk; AC: common carotid artery; ASUB: subclavian artery "string"; DAo: descending aorta.

the level of the origins of brachiocephalic and left common carotid arteries (Figs 7 and 8), producing a similar murmur that was also auscultated over the back.

Discussion

As far as we are aware, ours is the first description of tendinous structures in the aortic arch associated with innocent murmurs in childhood. There are, however, reports of such tendinous strings in the aorta following autopsy examination.

Such a tendinous string in the aortic arch was first described in 1912 by Luksch.[3] He reported a bow-like string 2 cm long and 1.5 cm thick, which extended across the lumen at the beginning of the ascending aorta. In 1936, Brenner[4] described an extraordinary and rare string-like structure in the ascending aorta of a 27-year-old accident victim. The cord was 2 cm long, 1.5 cm thick, was covered with intima, and coursed diagonally through the aortic lumen 4 cm above the arterial valve. At its endpoint, the string flattened and reached the aortic wall as a 2 mm broad ribbon. Histological examination showed surface endothelium covering concentric layers of material poor in nucleuses, but with many elastic fibres and smooth muscle tissue. The composition was similar to that expected from dissection of the aortic wall. Hill and

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Eismann[5] then observed a similar string in the aortic arch of a 65-year-old patient who died with myocardial infarction. The string reached across the lumen between the anterior and posterior aortic walls, being positioned between the brachiocephalic artery and the left common carotid artery.

It is understandable that such strings are usually missed on clinical examination. The murmurs we heard seemed innocent, so in many similar cases further diagnostic testing would be considered unnecessary. Our experience with 6 patients seen over a period of six months, nonetheless, suggests that the finding is not as rare as previously thought. The strings can be detected using cross-sectional echocardiography, as has also been shown in the diagnosis of tendinous ventricular cords. The images of the strings were seen by different examiners on different visits to the clinic.

As yet, there is no known pathological significance of these structures, and their aetiology is unknown, but murmurs are a frequent cause for referral to a paediatric cardiologist. Latest studies from Advani et al.[1] have shown that the clinical examination by an experienced paediatric cardiologist remains the most important investigation of all available diagnostic tests. Further testing is common in specialty centres, but rarely alters the final diagnosis.

In summary, over a short period of six months, we saw 6 patients in our outpatient clinic, each with fibrous strings crossing the aortic lumen, and each presenting with an innocent murmur. No history of cardiac dysfunction, cyanosis, syncope or arrhythmias could be obtained. All children were healthy, and were physically active. Apart from the murmur, physical examination was normal. We suggest that, in patients referred with innocent murmurs, if echocardiography is otherwise normal, and the murmur is inconsistent with Still's murmur or an aortic or pulmonary ejection murmur, then a thorough evaluation should be made to exclude the presence of tendinous structures in the aortic arch. The pathological significance of such strings, has yet to be determined, but we believe them to be the harmless cause of murmurs. More invasive diagnostic testing does not appear to be indicated. According to Smythe et al.,[6] even an echocardiogram is unnecessary if a diagnosis has been made on clinical examination by an experienced paediatric cardiologist.

"So, while listening let you guide yourself through the world of heart murmurs and you only see what you know".

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