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Letters to the Editor

Dear Sir,

We would like to draw your attention to another possible treatment for protein-losing enteropathy, a daunting problem following creation of the Fontan circulation.[1] We have been treating a 5-year-old boy, who had been a poor candidate for construction of the Fontan circulation, with a pulmonary vascular resistance of 4,7 WoodUm,[2] and mean pulmonary arterial pressure of 18 mmHg. He developed a protein-losing enteropathy about 4 months after the completion of a modified, fenestrated Fontan procedure at the age of one year. The enteropathy resolved after a pacemaker was implanted for treatment of sick sinus syndrome. Previous attempts using steroids or subcutaneous heparin failed to improve his enteropathy even in combination with a protein enriched diet.[2–4] The boy had an uneventful course until the fenestration of his caval tunnel was closed. His estimated capillary filtration pressure, according to a recently published algorithm, then increased to 20 mmHg.[5] About 8 weeks later, he again developed the protein losing enteropathy (Table 1). Explanting the device used to close the fenestration might have led to resolution of the enteropathy but would have meant further cardiac surgery.[6] After admission to our unit, we reduced his pacemaker settings from a minimum rate of 95/min at day-time and 85/min at night-time to 90/min respectively to 75/min, hoping this age-adjusted rate, in combination with a protein enriched diet, could improve his enteropathy. It did not improve (Table 1). The observations from Buchhorn et al.[7] suggested that beta-blockade decreased the capillary filtration pressure and elevated levels of renin and aldosterone. We reasoned that such therapy might also lead to resolution of the enteropathy. With the permission of his parents, we started with 2.5 mg metoprolol twice daily, increasing to 5 mg twice daily. This equated to a dosage of 0.5 mg/kg per day. All his symptoms resolved, though he started to complain of cold hands and feet (Table 1). We readjusted the dosing to 3 mg four times daily, equating to 0.7 mg/kg per day, and the coldness of his hands and feet improved. We tried twice to reduce his weight-adjusted metoprolol dosage, but twice this lead to diarrhea, which resolved as soon as the dosage of metoprolol was again increased. He has now been on a normal diet for more than 2 years, with a weight-adjusted dose of about 0.7 mg/kg per day. He remains free of any symptoms. The use of beta-blockade, therefore, is another option in patients with the Fontan circulation who suffer protein losing enteropathy. The question remaining is whether a fenestration should

Table 1. Symptoms, laboratory values and treatment in a boy with recurrent protein losing enteropathy.

Date	7/97	8/97	11/99	3/00	4/00	2/01	
Weight (kg)	14.1	13.4	17	19.1	18.4	18	
Stool	5	5	No	4	No	No	Diarrhea per day
Ascites	No	No	No	Yes	No	No	
α_1-antitrypsin	16	22		396	621		(<20) mg/100 g stool
Serum protein	3.5	5.8	5.9	3.3	4.3	6.9	(6.6–8.3) g/l
IgG	14.7	60.2		11.9	2.24		(7.25–18) g/l
IgA	3.3	2.8		2.7	4.9		(0.7–4.5) g/l
IgM	4.5	5.6		4.5	6.3		(0.6–2.8) g/l
Serum calcium	1.9	2.3	2.4	2.0	2.1	2.45	(2.1–2.8) mol/l
Renin				>25			(1.2–3.5) ngI/ml/h
Aldosterone				299	189		(30–150) ng/l
Therapy	Heparin	PM	Closure	PM-rate	Metoprolol		

Date: date of laboratory results; weight: body weight; stool: frequency of diarrhea, with "No" indicating one normal bowel motion per day; ascites: sonographic evidence of ascites; laboratory values of α_1-antitrypsin, serum protein, IgG, IgA, IgM, serum calcium, renin, and aldosterone in the blood with units and the normal values in brackets; therapy: the therapy initiated after obtaining the laboratory results; Heparin: heparin therapy; PM: implantation of a pacemaker; Closure: closure of the fenestration of the caval tunnel; PM-rate: reducing the pacemaker rate from 95 to 90/min during the day and 85 to 75/min at night and starting the metoprolol therapy; metoprolol: metoprolol to 3 mg four times daily increased

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not be closed in those who were initially poor candidates for establishment of the Fontan circulation?

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Dear Sir,

In the excellent paper by Nothroff and his colleagues,[1] recording the first clinical demonstration of the tendinous cords crossing the lumen of the aortic arch, it is stated that the aetiology of such structures is unknown. We believe that, in some cases at least, they may be explained by the minimal persistence of a fifth embryonic aortic arch. The most common form of this anomaly presents as a "double-barrelled", "double-lumen" or "subway" aortic arch, as first described by the Van Praaghs.[2] In 1989[3] we described a case in which the lumen of the aortic arch was crossed by a broad band which divided it into superior and inferior channels. Dissection showed that this band was not solid, but was a hollow tube, lined by adventitia which was in continuity with that of the aortic wall. It corresponded, therefore, to a "double-barrelled" aorta, with only a very short duplicated zone. We postulated that a more extensive coalescence of the walls of such a tube would produce a true band or cord across the aorta of the type described and illustrated by Hudson,[4] and similar to those illustrated by Nothroff and colleagues.[1] The ultimate degree of incorporation of the two channels would result in total disappearance of the cord. Such a happening would support the hypothesis that some apparently normal aortas may incorporate elements of the embryonic fifth aortic arch.

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Reply

The letter was shown to Dr Nothroff, who responded as follows:

Dear Sir,

I thank Drs Gerlis and Ho for their helpful comments. I have since discovered a few possible explanations for the aetiology of the strings. Brenner[1] described the string as an island of tissue between the fourth and fifth left branch arteries. He speculated that the island was formed by growth of the walls of the arteries of the fourth and fifth branchial arches as they combined to form a new common vessel. Luksch,[2] according to Henle, discussed a hindering in the melting process of the two embryogenic aortic roots, leading to the persistence of tendinous structures in the remaining vessel. To my knowledge, the aetiology of such strings has yet to be definitively proven. The hypothetic explanation provided by Gerlis and Ho will require further evidence from study of human embryos.

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