LETTERS TO THE EDITOR

Contractural arachnodactyly versus Marfan's syndrome

SIR,—I read with interest the case report by Huggon *et al* of an infant girl with contractural arachnodactyly, mitral regurgitation, and iridodonesis.¹ I agree that the presence of cardiovascular complications and iridodonesis obscures the differentiation between Marfan's syndrome and contractural arachnodactyly. However, there was no dilatation of the aortic root reported in their patient.

The most characteristic cardiovascular abnormality in Marfan's syndrome is aortic root dilatation.² Of all the cases of contractural arachnodactyly reported and cited as references by Huggon *et al*,¹ none had dilated aortic root. The only case quoted by the authors that had enlarged aortic root was diagnosed as having Marfan's syndrome with contractural arachnodactyly and severe mitral regurgitation.³

Although mitral valve prolapse has been found in association with Marfan's syndrome,⁴ the major cardiovascular lesion associated with this syndrome is dilatation of the root of the ascending aorta.² The latter is such a frequent (84%) finding,² far more than mitral valve prolapse (58%),² that in its absence one should seriously question the diagnosis of Marfan's syndrome.

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Growth failure secondary to moyamoya syndrome

SIR,—The case report by MacKenzie *et al*, which describes a 7 year old boy with growth failure secondary to moyamoya syndrome is interesting.¹ We would like to draw your readers' attention to a highly specific electroencephalographic finding in childhood moyamoya syndrome. This finding makes it possible to establish the tentative diagnosis of moyamoya syndrome on the basis of electroencephalographic recordings alone in many instances.

With childhood moyamoya syndrome the electroencephalographic tracing will show the so called rebuild up phenomenon after the hyperventilation test. This phenomenon is characterised by a renewed prolonged high amplitude frequency decrease in the electroencephalographic tracing after the hyperventilation test. It occurs only in association with childhood moyamoya syndrome, and is probably dependent on the degree of vascular change.

Electroencephalographic findings of 87 children with moyamoya syndrome have been reported in the literature. Only four children, who suffered from stage V moyamoya syndrome, did not develop any rebuild up phenomenon after the hyperventilation test.²⁻⁴ The rebuild up phenomenon might be attributed to increased vasoconstriction of the collateral vessels due to the induced decrease in oxygen, with delayed recovery after discontinuation of the hyperventilation test.

Our own finding of a rebuild up phenomenon in the electroencephalographic tracing, obtained in an 11 year old boy with no pertinent clinical symptoms, led us to establish the tentative diagnosis of a moyamoya syndrome which was confirmed by magnetic resonance imaging and angiography later.⁵

Regarding the patient described by MacKenzie *et al*, the moyamoya syndrome probably would have been diagnosed much earlier had the hyperventilation test been applied during the electroencephalographic recording and monitoring been continued sufficiently long. Due to the high specificity of the rebuild up phenomenon with regard to childhood moyamoya syndrome, we recommend magnetic resonance imaging or angiography whenever this electroencephalographic finding is obtained, even when clinical symptoms are absent. Only then will it be possible to extend proper advice to patients and parents.

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