more common in winter and worsen glucose control. Similarly, food intake is greater in the winter months when children gain relatively more weight than height. Seasonal variations in hormone concentrations may influence or be influenced by glucose concentrations.[4]

This study was done because of the clinical impression that diabetes was better controlled in the summer, and it is interesting that the mean amplitude of variation detected was small. This probably indicates that some patients have greater and therefore noticeable seasonal fluctuations; and it might be of clinical value to identify such children. This seasonal variation may be accentuated in higher latitudes where greater climatic differences between winter and summer occur.

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Mitral valve disease in Marfan's syndrome

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SUMMARY Cardiovascular disease in Marfan's syndrome presenting in childhood affects the mitral valve more often than the aortic valve or the aorta, as in adults. Early evaluation of the cardiovascular system is necessary for any child in whom Marfan's syndrome is suspected.

Marfan's syndrome comprises a characteristic phenotype with skeletal, ocular, and cardiovascular disorders. The dissection of the aorta and disorders of the aortic root which usually become apparent during the second and subsequent decades have been described. Impairment of the mitral valve is, however, also associated with Marfan's syndrome. Indeed, it is the most common cardiac manifestation in childhood and may present in the first year of life. We report on a child with Marfan's syndrome, which was apparent at birth, who died in early infancy from mitral valve disease. Early specialist evaluation of the cardiovascular system is essential in all cases of Marfan's syndrome.

Case report

The patient was the third child of unrelated white parents, with no family history to suggest Marfan's syndrome. Both siblings were well, the first having a

ventricular septal defect that had closed spontaneously. After a spontaneous labour at term delivery was vertex vaginal; birth weight was 4040 g (90th percentile), length 54·5 cm (0·7 cm above the 97th percentile), and occipitofrontal circumference 35·5 cm (90th percentile). Several abnormalities were apparent: the baby seemed abnormally long and slim with little subcutaneous fat; there was obvious arachnodactyly of both feet and hands (middle finger length 4 cm (97th percentile); metacarpal length was 1·9 cm (excluding the epiphysis; metacarpal index 8·25)) and span 57 cm (2·5 cm >length); she had a long thin face and sparse hair; the fingers and toes were hyperextensible; there was ulnar deviation of both hands and limitation of extension of both elbows and knees as well as pectus carinatum with prominence of the right side of the chest and divarication of the recti; and she had a high arched palate. A soft systolic murmur was audible at the first examination but subsequently disappeared.

Investigations in the neonatal period included chest radiography, electrocardiography, chromosome analysis, and estimation of plasma and urinary amino acid concentrations; all yielded normal results, except that the x-ray pictures showed the arachnodactyly. She was discharged home, being bottle fed, on the fourth day and followed up in the baby clinic at 7 and 12 weeks of age, when her
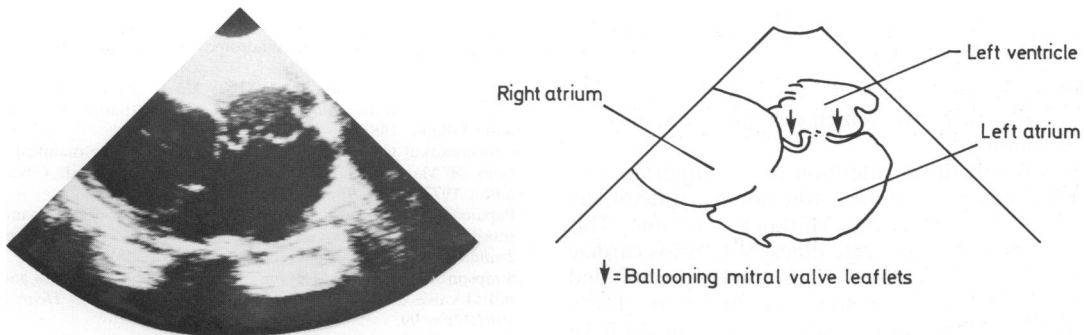
Figure   *Echocardiogram (foreshortened subcostal view) showing dilated left and right atria (LA, RA) and mitral leaflets billowing back into left atrium (arrowed).*

developmental progress was normal. Her weight gain was suboptimal, however, falling from the 90th percentile at birth to just below the 50th percentile at 12 weeks. Rate of growth for length and occipito-frontal circumference were normal. At the 12 week visit the soft systolic murmur was heard again.

At 16 weeks of age she was admitted severely ill with a two day history of fever, irritability, and tachypnoea. She had peripheral circulatory failure, central cyanosis, an active precordium with prominent ventricular impulses, and a loud systolic murmur; the liver could be felt 4 cm below the costal margin, and there were signs of left basal pneumonia. A chest *x*-ray film showed a large boot shaped heart with patchy shadowing in the right middle and left lower lobes. An electrocardiogram showed an axis of $+135°$ and right ventricular hypertrophy. Four hours after admission she collapsed after a convulsion and required intravenous fluid replacement and assisted ventilation.

An echocardiogram showed floppy prolapsing anterior and posterior mitral leaflets with a dilated valve ring (figure). All four chambers were dilated, as were the great vessels; injection of contrast showed severe tricuspid regurgitation, Full inotropic support was given together with diuretics and mannitol for oliguria. Asystole occurred 40 hours after admission, and she could not be resuscitated. Permission for necropsy was refused. The immediate family were subsequently screened; none had any suggestion of Marfan's syndrome or cardiac disease.

Discussion

In 1896 Marfan described a skeletal abnormality in a 5 year old girl characterised by abnormally long, slender extremities. The term arachnodactyly was suggested by Archard about six years later. Ironic-

ally, subsequent experience has cast doubt on whether this case was what we would today consider to be Marfan's syndrome with the characteristic phenotype and involvement of the skeletal, ophthalmological, and cardiovascular systems.[1] The syndrome is inherited in an autosomal dominant manner with variable expression, although about 15% of cases seem to be new mutations,[2] as this case.

Although the presentation of Marfan's syndrome in adults, with aortic dilatation, incompetence, or dissection, is well known, few paediatric text books make the point that in children and young adults with the disease mitral valve abnormalities are found more commonly than aortic lesions. Indeed, the most commonly used reference book about malformations fails even to mention that mitral valve disease may occur. Marfan's syndrome is rarely diagnosed in young children, and its full import needs to be emphasised.

Up to 61% of children with overt Marfan's syndrome may have appreciable heart disease. In one series of 36 children 12 girls and five boys had mitral regurgitation alone, three boys had mixed mitral and aortic regurgitation, and one boy had aortic regurgitation alone.[3] The aorta, aortic valve, sinuses of Valsalva, pulmonary artery, and mitral valve may all be affected.[3] [4] These abnormalities are part of a widespread connective tissue disorder affecting many organs. Although there may be accelerated linear growth in childhood, children with severe heart disease may fail to thrive. The gross signs in our patient may have implied more severe disease, and the failure to gain weight may have indicated severe cardiac disease.

Disease affecting the mitral valve is usually more benign than that of the aortic valve, but, rarely, acute emergencies may result from rupture of the chordae tendinae,[5] bacterial endocarditis,[6] or cardiac

failure precipitated by intercurrent infection, as in the patient reported on here. Surgical treatment for the floppy mitral valve abnormalities comprises repair or replacement of the valve. In young infants either of these carries a high risk and the results are disappointing.[3][5]

This report draws attention to the importance of careful assessment of the cardiovascular state of any child suspected of having Marfan's syndrome. This patient died after an acute illness with gross cardiac failure due to valve disease, presumably precipitated by an intercurrent respiratory infection. Early evaluation of the cardiovascular system might have resulted in earlier and possibly successful intervention.

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Bacterial tracheitis in Down's syndrome

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SUMMARY Four children with Down's syndrome and bacterial tracheitis are described. In three the infection was due to *Haemophilus influenzae*. In patients with Down's syndrome presenting with stridor tracheitis should be considered and appropriate treatment started.

Bacterial tracheitis (pseudomembranous croup) is characterised by upper airways obstruction with fever and is diagnosed by the presence on bronchoscopy of purulent tracheal secretions and a normal epiglottis. Most cases are due to *Staphylococcus aureus*, but *Haemophilus influenzae* and various streptococci have also been implicated.[1-4] Sofer *et al* reviewed 332 children with infective upper airways obstruction; 297 (89%) had croup, 28 (8%) epiglottitis, and 7 (2%) bacterial tracheitis.[2]

We report four children with Down's syndrome seen over three years with severe airways obstruction due to tracheitis. All were previously well and none had congenital heart disease. In three, *H. influenzae* was identified as the causative organism. Over the same period 206 children were seen with croup, and two with epiglottitis. None of them had Down's syndrome, nor was bacterial tracheitis diagnosed in any other child.

Case reports

Case 1. A 10 year old boy presented with a 24 hour history of cough, and a three hour history of increasing stridor. He was distressed, feverish, had a marked stridor, and showed signs of severe repiratory obstruction. Urgent bronchoscopy showed that the epiglottis was not enlarged and the larynx only slightly inflamed. The trachea was hyperaemic and oedematous, and copious mucopus was seen, which subsequently grew *H. influenzae*. Tracheal intubation relieved the obstruction and he breathed spontaneously. He was treated with ampicillin and flucloxicillin, tracheal aspiration, and physiotherapy. By the next day there was widespread consolidation of the lungs and enlargement of the mediastinum. His condition deteriorated and two days later ventilation was required. The antibiotics were changed to chloramphenicol and cefotaxime. With suction, tracheal lavage, and physiotherapy he gradually improved. He was extubated after seven days, and discharged well two days later.

Case 2. A 2½ year old boy was admitted with a six day history of measles. He had conjunctivitis, Koplik's spots, a morbilliform rash, cough, and mild stridor but no dyspnoea or recession. Three days after admission, over two to three hours he became