A new cardiomyopathy in girls

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SUMMARY A type of cardiomyopathy characterised by isolated severe dilatation of the right heart with right ventricular hypertrophy and absence of cardiac arrhythmias is described in seven patients. Only girls were affected and all except one lived in rural Zimbabwe. Six were under 5 years old. This probably represents a new type of cardiomyopathy.

Cardiomyopathies are uncommon in childhood but all the types seen in the adult population have also been described in children. Their overall contribution to childhood cardiac disease remains largely unknown. This article describes a form of cardiomyopathy that has so far not been described in children.

Patients and methods

Seven girls between 3 months and 14 years of age were seen over a 22 month period in our paediatric cardiac clinic for assessment of unexplained cardiomegaly or heart failure. Six were under the age of 5 years. All had chest radiographs and a 12 lead electrocardiogram was available in five patients.
Two dimensional echocardiography was performed using a 5 MHz Advanced Technology Laboratory Mark III sector scanner. Standard views were used for evaluation of cardiac anatomy.

**Results**

The clinical data on the seven girls are summarised in the table. Four patients presented with dyspnoea due to acute bronchopneumonia and two of these (cases 4 and 6) were in severe respiratory distress. In case 6 this was due to smoke inhalation and mild chest infection. One patient (case 1) was admitted with croup and developed bronchopneumonia and right heart failure two days later. Only two patients (cases 1 and 4) had previous hospital admissions, both for chest infections. The oldest patient, a 13 year old girl, had a history of exertional dyspnoea for two weeks and was initially thought to have rheumatic heart disease. No patient had clinical

<table>
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<th>Case no</th>
<th>Age</th>
<th>Presenting complaints</th>
<th>Cardiotoracic ratio</th>
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<td>Mean QRS axis</td>
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<tr>
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<td>2 years 8 months</td>
<td>Oedema, chest infection</td>
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**Figure** (a) Apical four chamber view of one of the girls showing a greatly dilated right heart and compressed small left chambers. The interatrial septum can be seen to be fully intact. (b) Two dimensional echocardiogram of one of the girls: the image is midway between apical four chamber and left parasternal long axis view. The aorta can be seen between the left atrium and the dilated right atrium. The right ventricle is very dilated and also hypertrophied and the left ventricle is compressed to a much smaller chamber. A, anterior; AO, aortic root; I, inferior; L, left; LA, left atrium; LV, left ventricle; P, posterior; R, right; RA, right atrium, RV, right ventricle; S, superior.
evidence of appreciable pulmonary hypertension. The precordium was remarkably quiet and left parasternal impulse was not prominent in any patient. The heart sounds were normal with the exception of one patient (case 7) with a slight accentuation of the pulmonary component of the second sound. A murmur of mild tricuspid regurgitation was present in only one patient (case 4). Right heart failure was present at some stage in all the patients. In cases 1 and 3 peripheral oedema was gross. History and clinical examination did not suggest cardiac arrhythmia in any patient.

The electrocardiogram showed sinus rhythm in all patients. The mean QRS axis ranged from 90–150° in four patients and was superior (−160°) in one. P wave was normal in all. Electrocardiographic evidence of right ventricular hypertrophy was present in three patients. The chest radiograph showed cardiomegaly in all and the most common pulmonary changes were pneumonic infiltrates, but two patients had completely normal lung fields.

The two dimensional echocardiographic features were identical in all the patients. The most striking abnormality was the severe dilatation of the right ventricle and right atrium (figure). The interatrial septum was intact in each case and could be seen bulging into the left atrium, thus reducing its cavity enormously. Pulmonary venous drainage was normal. The dilated right ventricle was slightly to moderately hypertrophied. The interventricular septum was intact and abnormal interventricular septal and ventricular wall motion was not prominent in any of the patients. The atrioventricular valves appeared normal and were normally located. The ventriculoarterial connections were concordant and no persistent ductus arteriosus was present. No patient had pericardial effusion.

Discussion

Harris and Nghiem defined cardiomyopathy in children as intrinsic disease of the myocardium that is not caused by shunts or valvar disease and subdivided them into primary and secondary types.1 The seven girls described here fulfil this definition and the cardiomyopathy can be regarded as primary. In children in developing countries the problem of recognition and classification of cardiomyopathies is often made difficult by the frequent occurrence of previously unrecognised rheumatic heart disease, which is quite common, uncorrected long standing congenital heart disease, and lack of adequate facilities and expertise for full investigation of cardiac disease. Many patients who died of cardiac disease will have had no necropsy and in cases where they were carried out the reports are often incomplete and unreliable. As rheumatic heart disease is common in developing countries there is a possibility of it being superimposed on a cardiomyopathy making recognition of the cardiomyopathy even more difficult. Most of the patients described here, however, are outside the usual age range for rheumatic heart disease.

Dilated cardiomyopathy is the commonest type of cardiomyopathy encountered in children and affects both ventricles with more severe involvement of the left side.2 It has a definite male preponderance and the first cases in children were reported from South Africa.3 4 Unlike the classic cases of dilated cardiomyopathy the seven children reported here have isolated right heart involvement. Other important features of these children are the exclusive involvement of girls and also their very young age.

The age of most of the patients would make one suspect congenital heart disease, notably total anomalous pulmonary venous drainage, as the cause of the cardiac problem. Venous drainage was, however, normal in each child and the obligatory atrial septal defect or patent foramen ovale of anomalous venous drainage was absent in all the girls. The possibility of primary pulmonary hypertension was excluded on clinical grounds. The fact that chest infection was common in these children does not necessarily mean left heart involvement as lower respiratory infections are extremely common in children in Zimbabwe and are by far the commonest cause of hospitalisation even in children without cardiac disease.

Only one patient was older than 5 years. The exclusive involvement of girls is unlikely to be a mere coincidence as biventricular dilated cardiomyopathy has been seen in both sexes in our child population. Another interesting and unexplained feature in these patients is the fact that all except the 3 month old infant lived in rural Zimbabwe.

A recent series from Hammersmith hospital reported on 14 cases of dilated cardiomyopathy with involvement of the right ventricle only.5 There was the usual male predominance in the series which included six children, four boys and two girls, the youngest of whom was 9 years old. The most prominent presenting features of those patients were syncope and cardiac arrhythmias, which were present in 70% of the cases. No patient in the present series had symptoms of cardiac irregularities and history and available electrocardiograms did not show any such disturbances. While there are similarities between the two groups of patients, there are obvious differences so that the children reported here most probably represent a different and new clinical entity.

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Diarrhoea caused by collagenous colitis

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SUMMARY The first case of collagenous colitis in a child with protracted watery diarrhoea and abdominal pain is reported. Small bowel investigations and the macroscopic appearances were normal, but histological examination of the colon showed collagenous colitis. Steroids temporarily relieved the diarrhoea and induced transient dissolution of the subepithelial collagen band.

Colitis in children is normally associated with bloody diarrhoea with mucus, tenesmus, and abdominal pain, and is usually caused by ulcerative colitis or Crohn's disease. Colitis caused by amoeba, campylobacter, antibiotics, food allergy, irradiation, or Hirschsprung's and Behçet's diseases are all uncommon. Recently microscopic and collagenous forms of colitis have been described in adults that cause profuse watery diarrhoea but remarkably occur without blood or mucus; thickening of the basement membrane is typical of this so called, 'collagenous' form. As far as we are aware no case has so far been reported in children, and we how report the first one.

Case report

A boy of 5 years old was referred to our unit with a four and a half year history of recurrent watery diarrhoea and abdominal pain, each episode lasting for about a week. During each episode his bowels opened up to 10 times a day, but he had otherwise grown and developed normally. His weight and height were on the 75th percentile, and between the 25th and the 50th percentiles, respectively. Examination showed no abnormal signs. Routine tests, including complete blood count, analysis of urine, plasma biochemistry, repeated stool culture and examination for parasites, stool chromato-graphy for sugars, sweat chloride excretion, faecal fat excretion, lactose and sucrose tolerance tests, small bowel biopsy, and barium follow through examination, were all normal; erythrocyte sedimentation rate was 29 mm in the first hour. Dietary exclusion of dairy products, eggs, fish, chocolate, sweets, fruit other than apples, and all processed foods, was of no benefit. Colonoscopy and ileoscopy showed no macroscopic abnormalities but histological examination of the biopsy specimens showed pronounced inflammation and thickening of the basement membrane.

HISTOPATHOLOGY

The initial biopsy specimens of large bowel mucosa were fixed in 10% formalin and embedded in paraffin; sections were then taken from several levels of each block and stained with haematoxylin and eosin, periodic acid-Schiff, van Gieson’s, and Congo red. The most striking abnormality was the increase in the thickening of the basement membrane between crypts. This stained pale pink with periodic acid-Schiff, and red with van Gieson’s, but did not show amyloid when stained with Congo red. One of us (GK) measured the basement membrane thickness using slides stained with haematoxylin and eosin and a Zeiss calibrated eye piece graticule. The thickness was estimated 30 times on each specimen in 25 adjacent intercrypt areas, and the mean thickness was 5-5 μm (range 3-7-9) (fig 1a). In addition the lamina propria was oedematous with an increased number of plasma cells and lymphocytes, particularly in the upper part of mucosa; there were no crypt abscesses or signs of ulceration.

In order to define the range of basement membrane thickness in children with other diarrhoeal diseases we retrospectively examined 543