ACUTE COR PULMONALE IN CHILDREN

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Chronic cor pulmonale has been observed frequently in children and has been reported most commonly as a complication of mucoviscidosis.[1] It has been described also in the following: tuberculosis, severe kyphoscoliosis, children suffering from primary vascular disease of the lung,[2-6] asthma,[9] and the Hamman-Rich syndrome.[7]

Acute cor pulmonale, on the other hand, has received little recognition as a complication of acute pulmonary conditions in childhood. The only unequivocal cases of acute cor pulmonale in children have been described by Royce,[1] viz. 2 fatalities caused by calcium chloride crystals in infused plasma. Controversial viewpoints are expressed on whether or not cardiac failure occurs as a complication of acute pneumonia. Royce[1] had seen a few babies, suffering from acute bronchiolitis, who had developed cardiac failure and who had recovered on digitalis therapy.

In an epidemic of acute bronchiolitis, Heycock and Noble[8] noted that the more severe cases, especially those with a fatal outcome, had shown signs of congestive cardiac failure. Similarly, Simpson,[9] in reviewing the case histories of 50 infants with heart failure, found 6 children in whom failure had been secondary to acute pulmonary infection. The Editor of the 'Year book of pediatrics',[10] who commented on this paper, doubted that heart failure was a complication of bronchiolitis and suggested that basal emphysema might be responsible for the engorged neck veins, the enlarged liver and the basal râles. Disney *et al.*,[11] commenting on an epidemic of bronchiolitis in infants, did not consider cardiac failure as a cause of death in their cases. The old concept of 'toxic' myocarditis complicating pneumonia appears to have been discarded.[12]

At Baragwanath Hospital we not infrequently encounter acute cor pulmonale as a complication of pneumonia in childhood. It is our purpose to describe the clinical features, the outcome and the treatment of this condition.

MATERIAL AND METHODS

This investigation is based on 178 consecutive Bantu children below the age of 9 years, who were admitted to 2 wards of the Baragwanath Hospital with pneumonia during the winter of 1958. X-ray examination of the chest was performed in all cases and electrocardiograms (ECGs) were obtained in 124 patients.

There were 155 children with bronchopneumonia. In the remaining 23 patients the bronchopneumonia was associated with pertussis. One child presenting with an acute pneumonia was later proved on lung biopsy to be suffering from the Hamman-Rich syndrome, but has been included in this series because of the acute mode of onset.

The patients were carefully examined for evidence of cor pulmonale; this was diagnosed in the presence of the following combination of signs: central cyanosis; increased venous pressure as shown by engorged neck veins and by enlargement of the liver; a hyperdynamic circulation characterized by a large pulse pressure, capillary pulsation and warm, moist extremities; clinical evidence of right ventricular dominance shown by a forceful left parasternal thrust; and the presence of a gallop rhythm. The ECGs were analysed for evidence of right ventricular hypertrophy and/or peaked P waves greater than 2·5 mm. in height.[13-15] Cardiomegaly was considered to be present on the X-ray film if the cardiothoracic ratio was greater than 55%.

Penicillin was the principal antibiotic used in the treatment of these children; usually 250,000 units were given every 6 hours by intramuscular injection. The antibiotic was changed if pyrexia had not subsided and the clinical condition of the patient had not improved within 72 hours of the commencement of therapy. If there was severe dyspnoea, cyanosis and restlessness, the child was placed in an oxygen tent.

Digoxin was given to 11 patients. The digitalizing dose was calculated on the basis of 0·08 mg. per kg. body weight, given in 3 equally divided doses by mouth at 8-hourly intervals. The maintenance dose consisted of $\frac{1}{6}$ - $\frac{1}{8}$ of the total digitalizing dose, and was given once daily. The drug was discontinued as soon as the signs of cardiac failure had disappeared.

RESULTS

There were 14 children who fulfilled the criteria for the diagnosis of cor pulmonale. Their ages ranged from 5 months to 4 years. One of the patients was 5 months old, 7 were aged 12 - 24 months and 6 were 25 - 48 months old.

Of the 14 children, 12 suffered from bronchopneumonia (4 cases being complications of pertussis), 1 suffered from giant-cell pneumonia, and the remaining 1 from the Hamman-Rich syndrome. Of the 4 children with pertussis bronchopneumonia, 1 had been whooping for 5 months, 2 for 2 months, and 1 for 3 days.

Central cyanosis was a well-marked feature in 12 of the 14 patients. In the remaining 2 children cyanosis was not easily recognizable because the children were anaemic. Their respective haemoglobin values were 8·5 and 8·8 G. per 100 ml. In contrast, only 14 (8%) of the remaining 164 patients with pneumonia were clinically cyanosed.

All the children with cor pulmonale had hyperdynamic circulations when first seen, as compared with 45 (27%) of the patients with uncomplicated pneumonia.

A presystolic gallop rhythm was found on auscultation in all 14 children. This finding was confirmed in 2 patients in whom phonocardiography was carried out. The disappearance of the gallop rhythm was the first sign of improvement in the 12 patients who recovered. The other 2 children died within 36 hours of admission to hospital, and the presystolic gallop rhythm persisted up to the time of death.

The 14 children with cor pulmonale were too ill for X-ray examination on admission to hospital, and this investigation was deferred until some improvement had been noted in their condition. Despite this delay, 8 of them showed radiological evidence of cardiomegaly. Furthermore, there was a return to normal heart size within 7 - 14 days in those patients who recovered.

No ECG abnormalities were detected in the patients with uncomplicated pneumonia. In the group with cor pulmonale, 8 (57%) showed evidence of right ventricular dominance, and of these 4 had enlarged, peaked P waves, i.e. P 'pulmonale'.

All 14 children received antibiotics and oxygen therapy, while 11 were given digoxin. On this therapy the gallop rhythm and the signs of cardiac failure disappeared within 24 hours in 5 patients and after 48 hours in a further 5. In 1 patient, who developed an empyema, signs of heart failure lasted for 4 days, and the patient suffering from the Hamman-Rich syndrome recovered from cardiac failure 7 days after admission to hospital.

There were 2 deaths. At autopsy, one child, aged 3 years, showed evidence of a giant-cell pneumonia and excess fat in the liver. The heart showed right ventricular hypertrophy and weighed 75 G. as compared with the average normal of 58 G. in this age group. The other child, aged 1½ years, had a severe necrotizing pneumonia. The heart, which showed evidence of right ventricular hypertrophy, weighed 67 G. as compared with an average normal weight of 52 G. at this age. There was no histological evidence of myocarditis in either of these children.

DISCUSSION

Our investigation shows that cor pulmonale is a not uncommon complication of severe acute pulmonary infections among children admitted to this hospital. This series does not give a true indication of the incidence of cor pulmonale, since many children suffering from pneumonia or whooping cough who attend at this hospital are treated as outpatients, and only the more seriously ill gain admission to the wards. Thus, an incidence of cor pulmonale of 7·8% in patients suffering from acute lung pathology refers only to the more seriously affected children.

The finding of a presystolic gallop rhythm in every patient with cor pulmonale requires comment. The additional heart sound is heard best just internal to the apical cardiac impulse and towards the left sternal border in the 4th intercostal space. It is an audible atrial sound.[16,17] So constant was this finding that we have included it among the diagnostic criteria for cor pulmonale. Furthermore, we found this sign to be of prognostic value, since its disappearance was followed by rapid clinical improvement.

It was of particular interest in 2 children. They were markedly dyspnoeic and cyanosed, with signs of heart failure and a presystolic gallop rhythm. They were given antibiotics and placed in oxygen tents. On examination the following day the signs of cardiac failure and the gallop rhythm had disappeared. When they were taken out of the oxygen tent, cyanosis reappeared and the gallop rhythm returned. With renewed administration of oxygen both the cyanosis and the gallop rhythm disappeared again. This phenomenon was experienced several times during the acute phase of the disease. These observations lend weight to the suggestion that anoxia (hypoxia) is the basis for the pulmonary hypertension, the increased cardiac output, and the cardiac failure in cor pulmonale.

The ECG and the X-ray examinations were of limited value in the diagnosis, since they showed right ventricular strain or cardiomegaly, respectively, in only half the patients.

Of the 23 patients with whooping-cough bronchopneumonia, 4 developed the clinical features of acute cor pulmonale. It would appear that the heart failure supervened in pertussis only when it was complicated by bronchopneumonia for, during the period under review, many patients with pertussis, unaccompanied by pneumonia, were seen in the outpatient department and none showed evidence of cor pulmonale.

Additional evidence that we were dealing with cor pulmonale was supplied by the autopsies performed on the 2 fatal cases. In both instances there was cardiac enlargement with right ventricular hypertrophy in association with pneumonia.

In the belief that hypoxia was the prime factor in the aetiology of our cases, we decided to omit digoxin in the last 3 patients and treat them with oxygen and antibiotics only. In these, and in 7 subsequent patients not included in this series, recovery from acute cor pulmonale

occurred within 48 hours of commencement of therapy. Thus, the prompt administration of oxygen is the single most important therapeutic measure in cases of acute cor pulmonale, even in the absence of clinical cyanosis, which may be masked by an underlying anaemia. From this experience it is suggested that digoxin be withheld unless signs of heart failure persist 48 hours after antibiotic and oxygen therapy is begun.

SUMMARY

Cor pulmonale is a well-described complication of chronic pulmonary disease, but is rarely mentioned as a complication of acute pulmonary infection in childhood. This series shows that, of 178 consecutive cases of severe pneumonia, there were 14 complicated by acute cor pulmonale. These included a patient with an acute onset of the Hamman-Rich syndrome.

The symptoms and signs of cor pulmonale in childhood are described, and special emphasis is placed on the development and prognostic significance of the presystolic gallop rhythm.

The clinical diagnosis of cardiopulmonary disease is emphasized as being more important and more accurate than either the ECG or radiological findings.

Autopsy evidence of right ventricular hypertrophy was found in 2 fatal cases of pneumonia and cor pulmonale. Children with pertussis may develop cardiac complications when there is an associated bronchopneumonia.

Hypoxia is the probable basis for the hyperdynamic circulation and the resultant cardiac failure. Prompt administration of oxygen was successful in overcoming right heart failure.

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