serum amylase level of 1400 IU/l and was thought probably to have had pancreatitis. It is concluded that while pancreatitis is an unusual cause of abdominal pain in childhood, it is common enough to warrant a serum amylase estimation if the pain is epigastric or if there is a known predisposing cause.

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References

- Adams, J. T., Elebute, E. A., and Schwartz, S. I. (1966). Isolated injury to the pancreas from non-penetrating trauma in children. *Journal of Trauma*, **6**, 86-98.
- Apley, J. (1975). *The Child with Abdominal Pains*, 2nd ed., p. 33. Blackwell, Oxford.
- Knight, A. H., Williams, D. N., Ellis, G., and Goldberg, D. M. (1973). Significance of hyperamylasaemia and abdominal pain in diabetic ketoacidosis. *British Medical Journal*, **3**, 128-131.
- Malone, J. I. (1974). Juvenile diabetes and acute pancreatitis. *Journal of Pediatrics*, **85**, 825-827.
- Shwachman, H., Lebanthal, E., and Khaw, K. T. (1975). Recurrent acute pancreatitis in patients with cystic fibrosis with normal pancreatic enzymes. *Pediatrics*, **55**, 86-95.
- Sibert, J. R. (1975). A British family with hereditary pancreatitis. *Gut*, **16**, 81-88.

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Influence of continuous positive airways pressure treatment on ductus arteriosus shunt assessed by echocardiography

In preterm infants the incidence of patent ductus arteriosus (PDA) is high. According to Kitterman *et al.* (1972) 15% of infants with a birthweight under 1750 g have clinical and haemodynamic evidence of PDA. The combination of respiratory distress syndrome (RDS) and PDA is often seen (Siassi *et al.*, 1969). By echocardiography it is possible to detect indirectly left-to-right shunting distal to the atrio-ventricular valves and to follow the effects of therapy on the shunt flow (Silverman *et al.*, 1974).

We present a case where decreased shunt flow during treatment with continuous positive airways pressure (CPAP) was demonstrable in an infant with RDS and PDA.

Case report

A male infant was born as the first of twins in the 28th gestational week with a birthweight of about 900 g; Apgar score was 1 and 5 at 1 and 5 minutes. He was ventilated for about 10 minutes, but then breathed spontaneously and had good colour. Both twins were immediately transferred to the Neonatal Intensive Care Unit.

During the first 24 hours he had apnoeic spells which increased in frequency. Chest x-ray showed a reticular-granular pattern suggestive of hyaline membrane disease. Treatment with CPAP applied with a face chamber was initiated and he improved. After 3 days of CPAP treatment he deteriorated with frequent apnoeic spells and intermittent positive pressure ventilation (IPPV) was necessary for 12 days. Thereafter he was weaned off IPPV and treated with CPAP only. In the following 4 weeks several attempts were made to take him off CPAP treatment, but each time he developed signs of cardiac decompensation with cyanosis, breathing difficulties, and hepatomegaly, signs that all disappeared as soon as CPAP was reinstituted.

At one week of age a systolic murmur was heard and repeat chest x-rays showed increased cardiac size and dilated pulmonary arteries. Digitalis treatment was started when he was 3 weeks old. At 41 days of age the systolic murmur disappeared and CPAP and digitalis treatment could finally be discontinued. After this his course was uneventful; chest x-ray normalized, he gained in weight, and was discharged aged 13 weeks. At age 4 months psychomotor development was normal.

Echocardiography. Echocardiography employed a technique previously described (Lundström, 1974) using a Smith-Kline Ekoline 20 Ultrasonoscope and a 5 MHz unfocused transducer. Polaroid photographs of the time-motion display were taken from a slave storage oscilloscope (Tektronix 603). On the echocardiogram measurements were made of the aortic root diameter (Ao) and the left atrial dimension (LA) at end systole, defined as the point of maximal forward movement of the aortic root (Fig. 1). The ratio LA/Ao was used to assess left atrial size, and thereby indirectly shunt flow (Silverman *et al.*, 1974).

In a group of 34 premature infants without clinical or x-ray evidence of cardiac disease, 54 measurements of LA/Ao ratio were performed with consecutive examinations at different weight levels. For this normal group we found a mean LA/Ao ratio of 1.12 ± 0.28 (2 SD).

Our patient was examined by echocardiography initially at the age of 3 days, again when the systolic murmur appeared, and then several times during the

Fig. 1 Echocardiographic examination of the aortic root and left atrium. (A) Route of the sonic beam as it traverses aortic root and left atrium. (B) Echocardiogram, sites for measurement of aortic root diameter (Ao) and left atrial dimension (LA) are shown.

Fig. 2 (A) Changes in LA/Ao ratio. Periods of IPPV and CPAP are indicated. The ratio increases when CPAP is withdrawn, decreases when CPAP is reinstituted, and returns to normal after spontaneous closure of the PDA. (B) Enlarged areas showing the rapid changes in LA/Ao ratio with and without CPAP treatment.

critical period. We observed that an increased LA/Ao ratio correlated well with clinical and x-ray evidence of increasing shunt and cardiac decompensation. During CPAP face chamber treatment the LA/Ao ratio decreased. As soon as CPAP was discontinued the ratio immediately increased but returned to 'treatment levels' when CPAP was reinstituted (Fig. 2). When the systolic murmur disappeared the ratio

normalized, and at follow-up at 8 and 11 weeks the values were within normal range.

Discussion

in some infants with RDS plus PDA there is an increasing left-to-right shunt which can result in congestive heart failure and pulmonary oedema. In

such situations IPPV has been used for combating severe oedema (Robin *et al.*, 1973). In other reports surgical ligation of the patent ductus has been advocated (Gay *et al.*, 1973). However, treatment with CPAP for prolonged periods can be a most efficient therapy as shown in our case. In our experience, as in Robertson's (1974), surgical treatment can be avoided and IPPV is only necessary in the most severe cases for a short time, followed by CPAP.

Transpulmonary distending pressure with CPAP has become an established method for treatment of RDS. CPAP via face chamber (Fc 100, Siemens-Elcoma) has been used for prolonged periods without hazards (Ahlström *et al.*, 1976). The cardiopulmonary effects of an increased transpulmonary pressure include counteraction of alveolar collapse with opening of atelectases, reducing intrapulmonary shunting, and increasing oxygenation. The resulting increase in arterial oxygenation presumably initiates ductal constriction. In addition the applied pressure may have some effect on concomitant pulmonary oedema.

Our observations show that changes in left atrial size, indicating changes in shunt flow even within a short period of time, can be followed closely by repeated echocardiographic examination. In this way the duration of CPAP treatment for infants with RDS plus PDA can be decided.

Summary

In a preterm infant with the respiratory distress syndrome complicated by patent ductus arteriosus, continuous positive airways pressure (CPAP) treatment relieved the signs of cardiac decompensation associated with left-to-right shunt. Echocardiography enabled the change in left atrial size, an indirect measure of the shunt, to be followed. In this way the rapid effect of CPAP in reducing left-to-right shunting could be monitored. This noninvasive technique could have many applications in neonatology.

References

- Ahlström, H., Jonson, B., and Svenningsen, N. W. (1976). Continuous positive airways pressure treatment by a face chamber in idiopathic respiratory distress syndrome. *Archives of Disease in Childhood*, **51**, 13–21.
- Gay, J. H., Daily, W. J. R., Meyer, B. H. P., Trump, D. S., Cloud, D. T., and Molthan, M. E. (1973). Ligation of the patent ductus arteriosus in premature infants. Report of 45 cases. *Journal of Pediatric Surgery*, **8**, 677–683.
- Kitterman, J. A., Edmunds, L. H., Jr., Gregory, G. A., Heymann, M. A., Tooley, W. H., and Rudolph, A. M. (1972). Patent ductus arteriosus in premature infants. Incidence, relation to pulmonary disease and management. *New England Journal of Medicine*, **287**, 473–477.
- Lundström, N.-R. (1974). Clinical applications of echocardiography in infants and children. *Acta Paediatrica Scandinavica*, Suppl. **243**.
- Robertson, N. R. C. (1974). Prolonged continuous positive airways pressure for pulmonary oedema due to persistent ductus arteriosus in the newborn. *Archives of Disease in Childhood*, **49**, 585–587.
- Robin, E., Cross, C. E., and Zelis, R. (1973). Pulmonary edema. *New England Journal of Medicine*, **288**, 292–304.
- Siassi, B., Emmanouilides, G. C., Cleveland, R. J., and Hirose, F. (1969). Patent ductus arteriosus complicating prolonged assisted ventilation in respiratory distress syndrome. *Journal of Pediatrics*, **74**, 11–19.
- Silverman, N. H., Lewis, A. B., Heymann, M. A., and Rudolph, A. M. (1974). Echocardiographic assessment of ductus arteriosus shunt in premature infants. *Circulation*, **50**, 821–825.

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Cystinotic rickets treated with vitamin D metabolites

Nephropathic cystinosis is a lethal inborn error of metabolism characterized by the autosomal recessive inheritance of an unknown biochemical defect which produces massive intracellular cystine accumulation. The disorder presents in childhood with failure to thrive, thirst, dehydration, stunting of growth, photophobia, and rickets. Most of these features appear to be secondary to a widespread failure of renal tubular reabsorptive function, which produces glycosuria, phosphaturia, generalized aminoaciduria, renal tubular acidosis, and hypokalaemia (Schneider and Seegmiller, 1972). The rickets may have several possible causes which include acidosis, hypophosphataemia, and renal glomerular failure. Treatment of the bone disease usually involves correction of the acidosis, together with administration of vitamin D in large doses and of phosphate, separately or in combination.

The rickets of cystinosis does not respond to physiological doses of vitamin D and part of this resistance may be due to reduced formation of the active metabolite 1,25 dihydroxycholecalciferol (1,25-DHCC) (Kodicek, 1974) in the diseased kidney.

In short-term studies, Balsan *et al.* (1975) were unable to detect any biochemical change in 2 cystinotic children given 2 µg 1,25-DHCC in daily doses, but Gertner *et al.* (1976) demonstrated healing of rickets in 3 boys with cystinosis given 1α-hydroxy-