in which there was rapidly progressive unilateral exophthalmos. In the report of Galyean and Robertson (1970), the patient presented in much the same way as ours with puffy eyes, irritability, and poor feeding. This child had glaucoma which resolved along with the hyperostosis over a period of 4 months.

The occurrence of thrombocytosis in Caffey's disease is now well recognized. Pickering and Cuddigan (1969) described 3 cases, all with raised platelet counts. In their review of reports published in English they found 3 cases with platelet counts $> 500\,000/\text{mm}^3$, this measurement having been made in only 9 of 132 cases. Our patient's platelets rose to $910\,000/\text{mm}^3$ but she suffered no thrombotic complications and no anticoagulant or antimegakaryocytic agents were used.

The reason for the thrombocytosis in Caffey's disease is not known. The low vitamin E levels shown in our patient may be a fortuitous finding but it is tempting to postulate a possible association between the thrombocytosis and the vitamin E deficiency in our patient. Ritchie *et al.* (1968) described a syndrome of oedema, haemolytic anaemia, and thrombocytosis in association with vitamin E deficiency. Thrombocytosis was also noted by Hassan *et al.* (1966) in their description of a syndrome in premature infants associated with low plasma vitamin E levels and a high polyunsaturated fatty acid diet. Our patient was fed on Cow and Gate 'Premium', then changed to Ostermilk II. She received Abidec drops (vitamins A, B, C, D) but no vitamin E. The plasma vitamin E level rose spontaneously to reach the lower level of normal after 3 months. There was no evidence of haemolysis.

Whether vitamin E deficiency is a factor in the pathogenesis of Caffey's disease remains to be seen. It is hoped that this report will stimulate further studies of vitamin E levels in infantile cortical hyperostosis.

Summary

An infant is described who presented with ptosis and periorbital oedema and was found to have infantile cortical hyperostosis with thrombocytosis, raised IgM, and vitamin E deficiency.

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Gastric pneumatosis in infancy

Gastric pneumatosis (defined as gas within the wall of the stomach) may be easily detected by radiography. It presents as a fine lucent stripe conforming to the contour of the stomach and enveloping any intraluminal gas and fluid content. The finding, though quite rare, is of utmost clinical importance. In adults, gastric pneumatosis is caused either by gas-forming bacterial infection (emphysematous gastritis), or in association with bullous emphysema (Holgersen, Borns, and Srouji, 1974). In infancy, isolated gastric pneumatosis has been seen very rarely in gastric outlet obstruction. It has also been reported in neonatal necrotizing enterocolitis, in association with intestinal pneumatosis (Bell, Graham, and Stevenson, 1971; Santulli *et al.*, 1975) though it may rarely be isolated (Robinson, Grossman, and Brumley, 1974). Thus, radiographic detection of gastric pneumatosis indicates serious underlying disease and determination of its cause will depend on the associated clinical findings, a point illustrated by the 2 cases presented here.

Case reports

Case 1. A 6-week-old girl developed vomiting, constipation, and oliguria 5 days before admission. She was a well-nourished infant, mildly dehydrated, and irritable. There was moderate epigastric distension but no visible peristaltic waves. No pyloric 'tumour' could be palpated. Laboratory studies including serum electrolytes were normal, except for slightly raised blood urea nitrogen ($18.7\text{ mg}/100\text{ ml}$). Radiographic examination of the abdomen showed gastric distension with a long air fluid level in the stomach seen on the upright film. The findings suggested gastric retention.

FIG. 1.—Case 1. Upright film. Gastric distension; intramural air around the fluid-filled gastric antrum.

A continuous curvilinear lucent stripe was noted to envelop the gastric contour, typical of intramural gas

(gastric pneumatosis) (Fig. 1). Small amounts of gas were scattered in the small bowel and colon.

FIG. 2.—Case 2. Linear pneumatosis surrounds the air-filled fundus of the stomach (arrows).

The clinical and radiological findings led to the diagnosis of gastric outlet obstruction. A nasogastric catheter was passed into the stomach and copious amounts of gastric contents were aspirated. Within a few hours a second radiographic examination showed marked gastric decompression and disappearance of gastric pneumatosis. Barium introduced in the stomach via the catheter under fluoroscopic control showed the typical x-ray signs of hypertrophic pyloric stenosis. At operation a long leathery pyloric sphincter was found and pyloromyotomy carried out. The patient tolerated the procedure well and has remained well.

Case 2. A female infant was born to a 21-year-old gravida 2, para 3 mother, after approximately 28 weeks' gestation. She was the second of twins and weighed 1178 g. Dyspnoea, tachypnoea, expiratory grunt, and substernal retractions were noted soon after birth, and on auscultation of the lungs there were fine crackling râles bilaterally. An umbilical artery catheter was

placed in the mid-thoracic aorta. Blood gases gave pH 7.23, PaO_2 70 mmHg, and PaCO_2 36 mmHg in $\text{F}_{\text{I}}\text{O}_2$ 30%. A chest x-ray was consistent with mild idiopathic respiratory distress syndrome. She responded well to moderately increased concentration of inspired O_2 administered by continuous positive airway pressure. A right upper lobe pneumonia was identified on chest x-rays on the 13th day and tracheal aspirate grew pseudomonas. She was treated successfully with carbenicillin and gentamicin. Her course was also complicated by hyperbilirubinaemia and a persistent ductus arteriosus for which she was treated with digitalis.

A transpyloric tube was used for nasojejunal feedings on the 15th day but was discontinued 2 days later because of abdominal distension. Diarrhoea, recurrent distension, and diminished bowel sounds were noted on the 33rd day. Abdominal films showed gastric pneumatosis (Fig. 2) and colonic dilatation, but there was no evidence of intestinal pneumatosis. Oral feedings were discontinued, intravenous fluids were started, and she

was given penicillin and gentamicin for 5 days. During this time abdominal distension subsided, bowel sounds became active, and diarrhoea decreased. Gastric pneumatosis disappeared 2 days after onset of therapy. The infant had a protracted course with poor weight gain and periods of apnoea and bradycardia. She was finally discharged at the age of 3½ months in good health.

Discussion

X-ray diagnosis of gastric outlet obstruction in infancy is very difficult without contrast media. Certain plain film signs are suggestive, but not diagnostic (Riggs and Long, 1971). Although gastric pneumatosis is a very rare radiographic manifestation of pyloric obstruction (e.g. from hypertrophic pyloric stenosis) it should immediately lead to the correct diagnosis, provided that sepsis and necrotizing enterocolitis can be excluded, readily accomplished on clinical grounds. Necrotizing enterocolitis is seen among preterm infants, usually after perinatal stress. There are signs of sepsis, abdominal distension, decreased or absent bowel sounds, bile-stained vomiting, and often bloody stools. In addition, in most cases there is radiographic evidence of generalized bowel distension and intestinal pneumatosis, though occasionally necrotizing or ischaemic gastroenterocolitis may present radiographically by isolated gastric pneumatosis, as in Case 2.

Depending on the differential diagnosis described above, infants with suspected gastric outlet obstruction should have a barium upper gastrointestinal study to show the exact nature of obstruction. Sepsis and ischaemic gastroenterocolitis should be treated immediately and vigorously as outlined by Santuli *et al.* (1975).

Speculating on the mechanism of gastric pneumatosis gastric outlet obstruction may lead to increased intraluminal pressure and gas may be forced into the wall of the stomach through superficial mucosal tears (Holgersen *et al.*, 1974). Disappearance of gastric pneumatosis in our Case 1 within hours after gastric decompression was in keeping with this. Mucosal tears are likely to occur with excessive vomiting, and indeed 3 infants described by Holgersen *et al.* (1974) had haematemesis or coffee-ground vomitus. In necrotizing enterocolitis presumably

breakdown of effective mucosal barrier leads to invasion by gas-forming bacteria, which may then produce intramural gas (Stone *et al.*, 1968). Regarding our Case 2, presumably she had emphysematous gastritis, as a form of ischaemic gastroenterocolitis of the newborn.

Summary

Gastric pneumatosis (gas in the wall of the stomach) is an uncommon but characteristic plain-film radiographic sign. In infancy it is associated either with gastric outlet obstruction, usually hypertrophic pyloric stenosis, or necrotizing gastroenterocolitis. Differential diagnosis is mainly based on associated clinical findings, as exemplified by the 2 cases reported here. Both infants had isolated gastric pneumatosis as the principal radiographic finding. Correct diagnosis was greatly aided by recognition of this x-ray sign so that appropriate therapy was started without delay.

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