did not investigate the reasons for non-compliance. It is not possible that the guardians were too sick with their pulmonary tuberculosis or HIV (not mentioned in the study, but assumed to be endemic in the suburbs of South Africa) to bring their children regularly to the clinics. On the basis of anecdotal experience of non-compliance, we use dual therapy with rifampicin and isoniazid. As soon as any non-compliers are located, intensive supervised treatment or directly observed therapy is instigated.

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Chronic intestinal ischaemia and Hirschsprung's disease

The prolonged carriage of *Clostridium difficile* in Hirschsprung's disease reported in this study¹ raises the possibility that Hirschsprung's might indeed be a product of intestinal ischaemia that develops in utero and might persist thereafter as chronic intestinal ischaemia, possibly because of persistent vascular abnormalities.²

Chronic ileal ischaemia seems to be the primary cause of pouchitis and the septic complications and anastomotic leaks complicating restorative proctocolectomy of ulcerative colitis.³ Anaerobes proliferate in a newly constructed pouch,⁴ and when pouchitis develops, these may include *C. difficile*. This is consistent with the proposal that *C. difficile* might be a marker of ischaemic colitis.⁵ Pseudomembranous enterocolitis and haemorrhagic necrotising enterocolitis complicating Hirschsprung's might also be products of intestinal ischaemia.⁶

Objective evidence of chronic intestinal ischaemia might only be apparent in a stress test, notably gastric exercise tonometry in the adult.⁷ If, however, histopathic hypoxia is a contributing factor, as is likely with any inflammatory disease superimposed on the Hirschsprung's, it should be evident from tonometric measurements performed at rest. It would be advisable to include the intramucosal pH as an end point in performing tonometry⁸; the partial pressure of the carbon dioxide-gap may not be increased in these circumstances.⁹ From our experience with chronic gastric ischaemia, enteral feeding even with cold water might be conveniently used as a stress test in infants. This may not be without risk in the critically ill in whom histotoxic or cytopathic hypoxia might be present.^{10–11}

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Trisomy 13 is associated with anorectal malformations

Babies with trisomy 13 (Patau's syndrome) have a median survival of 2.5 days, with only 5% surviving beyond 6 months. There are three cases of trisomy 13 with anorectal malformation in the English literature; a partial trisomy of chromosome 13q and 20p,¹ a mosaic form (46,XX,t(13q13q)/46,XX,-13,+r(13))² of trisomy 13 and a third case in which the form of trisomy 13 was not described.³ We report a case of full trisomy 13 with an unusual anorectal malformation.

A boy delivered at 36 weeks' gestation had antenatal scans showing a cleft lip and a large ventricular septal defect with an over-riding aorta. He was below the third centile for weight and head circumference. He had a small occiput with occipital and parietal scalp defects, micro-ophthalmia, a cleft lip and palate, fistled hands, rocker-bottom feet,

micropenis, an impalpable right testis and an imperforate anus. Echocardiography showed a patent ductus arteriosus and coarctation of the descending aorta in addition to a ventricular septal defect and an over-riding aorta. Fluorescent *in situ* hybridisation and karyotype analysis confirmed trisomy 13 with a Robertsonian translocation of the extra chromosome 13 to chromosome 14.

On day 2, he had increasing abdominal distension and pain. A plain abdominal film showed a hugely distended colon with no gas beyond the pelvic brim (fig 1). Importantly, the gas pattern in the sigmoid colon was limited to the left iliac fossa, whereas distension of the sigmoid typically displaces the sigmoid colon to the right in imperforate anus.

Despite the short life expectancy, we performed a colostomy under general anaesthesia for the palliative relief of intestinal obstruction at 64 h. At laparotomy, his distended sigmoid colon had no mesentery, and it was difficult to mobilise. This is unusual. A flush colostomy was therefore formed in the anterior wall of the sigmoid colon. He was comfortable post-operatively, but developed increasing respiratory distress and died at 84 h. No postmortem examination was performed, and we do not know whether there was a fistula to the urinary tract.

Interestingly, in a study of 1992 patients with anorectal malformation, 5% had trisomy 21 but none had trisomy 13.⁴ Of the 1992 patients, 238 had no fistula and 40% of patients without fistulas had trisomy 21. In a smaller study of 103 patients with anorectal malformation, one had trisomy 13.⁵ Trisomy 13 with a coexisting anorectal malformation could be a chance occurrence. Alternatively, a genuine association between trisomy 13 and anorectal malformations exists, and the unusual anorectal anomaly in this case supports this hypothesis.

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