normal amount of meconium in the first few days of life, with fairly diffuse distribution of gas on earlier x-ray films (Fig.). Signs of duodenal obstruction with paucity of gas distally became evident on later films. Not surprisingly, the operative manoeuvres were aimed at the duodenum, whereas in the previously reported cases the common operation performed was either a colostomy or an ileostomy.

Malrotation of the intestines has been reported previously in association with Hirschsprung's disease (Rickham and Johnston, 1969) and has been seen in other cases of Hirschsprung's disease in Manchester (S. Ahmed, unpublished). The presence of malrotation complicated the clinical picture in this case. It appears that in cases of malrotation or when the exact nature of the duodenal obstruction is not clear, the appendix should be taken out for urgent histological examination. If Hirschsprung's disease is diagnosed, further serial biopsies of the ileum and jejunum should always be undertaken as location of the cone area is notoriously difficult in such cases. The degree of involvement of the small bowel would thus be elucidated, enabling one to perform the appropriate surgical procedure. On the other hand, knowledge of the presence of total intestinal aganglionosis would contraindicate further surgical measures, the condition being incompatible with life.

**Summary**

A case of total intestinal aganglionosis is reported. The patient presented as a duodenal obstruction with intestinal malrotation. The reason for the failure of operative manoeuvres to relieve the obstruction was suspected before the patient died, and confirmed on histological studies which showed absence of ganglion cells and nerve fibres from the anal canal to the duodenum.

We would like to thank Dr. John Fawcitt for radiological comments and Miss Perry for the illustration.

**REFERENCES**


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**Dosage of Oral Salbutamol in Asthmatic Children**

Salbutamol has been shown to be a potent bronchodilator with less cardiac action than isoprenaline (Warrell et al., 1970; Kamburoff and Prime, 1970) or orciprenaline (Riding, Dinda, and Chatterjee, 1970). This is thought to be due to its selective activity on β-2 adrenergic receptors in bronchial muscle (Cullum et al., 1969). The optimal dose for use clinically should reduce airways resistance without causing side effects, particularly tachycardia. Therefore a dose response study was undertaken to select a dose of oral salbutamol which would give maximal relief to a child with
asthma (judged by a rise in peak expiratory flow rate) without causing a significant increase in heart-rate.

Method

Sixteen asthmatic children, aged between 6 and 13 years, of both sexes, were tested with a placebo and salbutamol syrup; a dose of 0.05 mg or 0.1 mg or 0.15 mg/kg was used. The children were all known to have previously responded to a sympathomimetic bronchodilator but had not received any treatment in the preceding 12 hours. None was taking steroids. On the day of the study they had resting peak expiratory flows (PEF) of less than 65% of predicted and were given one syrup in the morning and the other in the afternoon, in random order and in a double-blind fashion. For each test a basal record of PEF (the maximum of triplicate readings) and of the radial pulse over 1 minute was made and thereafter followed up serially for 3 hours. During this period the children were kept as quiet as possible.

Results and Conclusions

The results, i.e. changes in PEF and pulse rate during the study, have been expressed as a percentage increase or decrease from the resting level at half an hour and 1, 2, and 3 hours. They are shown in Fig. 1–3 as the mean change for each dose group and corresponding placebo study. All doses of salbutamol produced a significant improvement in PEF not seen when the placebo was taken. The effect on PEF took place within half an hour, with maximal improvement occurring at 2 to 3 hours; however the children were not followed up for a longer period. No group showed a significantly greater rise of pulse rate after salbutamol than after placebo syrup.
Salbutamol syrup (0·05 mg–0·15 mg) was therefore effective in improving the PEF in the presence of bronchospasm in this group of children with mild/moderate asthma. As the greatest improvement in PEF was seen in the group taking 0·15 mg/kg without any side effects or rise in pulse rate, this seems to be the dose of choice.

Summary
Salbutamol syrup (0·05 mg or 0·1 mg or 0·15 mg/kg) was tested on 16 asthmatic children. Placebo syrup was used as a control and the effect on the peak expiratory flow and pulse rate was noted for the next 3 hours. The highest dose was shown to be the most effective in increasing the PEF without causing a rise in the pulse rate.

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A-β-lipoproteinaemia and Colour-blindness

A-β-lipoproteinaemia is a hereditary disorder characterized by the absence plasma of low and very low density lipoproteins as well as chylomicra (Kahlke, 1967). Clinically the disease is manifested by steatorrhoea and failure to thrive, progressive ataxic neuropathy, pigmentary changes of the retina, and acanthocytosis. The present report describes the disease in an Arab boy in whom the condition was associated with colour blindness.

Case Report
A 15-year-old Arab Jordanian boy was admitted to the American University Hospital for the investigation of his ataxic gait. Fat intolerance had been present since infancy and colour blindness responding to vitamin A was noted at age 2. Muscle weakness developed at age 7 followed by ataxia and deterioration of performance at school.

The parents were first cousins and had 3 older unaffected boys. The father had bilateral xanthelasma and a plasma cholesterol of 287 mg/100 ml. The mother’s plasma cholesterol was 235 mg/100 ml.

He was a thin boy who looked younger than his age and walked with a wide-base ataxic gait. His height was 140 cm and his weight 31 kg. The upper teeth were protruding outwards and there was inward curving of the fourth and fifth left fingers with a swan neck appearance. The genitalia were small and the pubic and axillary hair were infantile. The cranial nerves were intact. There was fine tremor of the tongue. The deep tendon reflexes were absent and the superficial reflexes were depressed. Vibration and position sense were absent; the finger-to-nose, heel-to-shin, and Romberg tests were all positive. The psychological test (WISC) (Dr. U. S. Yaktin) showed a verbal IQ of 90, performance IQ of 63, and full-scale IQ of 72. His poor performance was ascribed to perceptual difficulties related to his physical condition rather than to mental retardation.

The significant ophthalmological findings were: bilateral myopia of 3-50 dioptres was discovered by cycloplegic refraction, and the best corrected visual acuity was 20/40 O.U. The ocular media were clear. An alternating exotropia of 20 prism dioptres was present. The ophthalmoscopic findings were similar in both eyes; there was a small temporal myopic crescent of the optic disc; the foveal reflex was absent; the

![](image_url)

**Fig. 1.** Right macular area. Note tessellated appearance and areas of discrete pigment loss.