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 of 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-based 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 gentalia 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 3–5 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

![Fig. 1.—Right macular area. Note tessellated appearance and areas of discrete pigment loss.](http://adc.bmj.com/10.1136/adc.46.250.871)
retinal vessels appeared normal; the posterior pole was heavily pigmented compared to the periphery of the retina and had a tessellated appearance (Fig. 1); posterior and peripheral small areas of discrete pigment loss were present; there were a few linear areas of pigment clumping at the periphery (Fig. 2). Goldmann perimetry showed constriction of the peripheral visual fields O.U. to 30 and 50 degrees with the I/4 and II/4 white isopters, respectively. The colour vision tested with the H-R-R pseudoisochromatic plates showed a strong blue-yellow defect. Dark adaptation, by the Bausch and Lomb modified orthorater, showed complete absence of dark adaptation within 30 minutes and subnormal post-adaptation acuity (Omarzu, 1970; Haddad, 1970).

The ophthalmological examination of both parents and of the oldest brother, including visual acuity, ophthalmoscopy, refraction, and colour vision, did not reveal any abnormal findings.

The red cells showed no sedimentation after 1 hour. Peripheral smear showed marked acanthocytosis, more marked on the centre of the smear. Immunoelectrophoresis of serum proteins showed normal pattern except for marked decrease of IgM. Plasma cholesterol ranged between 36 and 40 mg/100 ml on eight occasions, phospholipid phosphorus 2-1, and triglycerides 6 mg/100 ml. Paper electrophoresis, immunoelectrophoresis, and ultracentrifugation revealed complete absence of β-lipoproteins, the α-lipoprotein accounting for the total plasma lipids.

Serum vitamin A level was 47 μg/100 ml (normal 30-50 μg/100 ml) and the serum carotene level was not measurable.

Vitamin A administered intramuscularly over a four-month period failed to raise the plasma vitamin A level or affect the ophthalmological examination. When the patient was given a diet in which 50% of the calories were derived from a medium-chain triglyceride containing milk (Portagen, Mead Johnson Co.), there was a decrease in the bowel movements, improvement in the upper gastrointestinal series and in muscle tone, but there was no improvement in the neurological findings.

Discussion

Approximately 30 cases of α-β-lipoproteinaemia have been reported in the world literature, but this is the first report of the condition in an Arab. The analysis of existing pedigrees as well as ours is consistent with the hypothesis that the condition is transmitted by a single autosomal recessive gene. The hyper-β-lipoproteinaemia found in the father of our patient has also been reported by Forsyth, Lloyd, and Fosbrooke (1965).

Pigmentary degeneration of the retina, night-blindness, constriction of visual fields, and exotropia have been previously reported in α-β-lipoproteinaemia (Kornzweig, 1970). In addition to these abnormalities, our patient exhibited defective blue-yellow colour vision or tritanopia.

Tritanopia is one of the rare types of colour-blindness. It is inherited as an autosomal dominant gene which may have variable penetrance, but X-linked transmission of the defect may also occur in rare instances (Kalmus, 1955). Since the parents and one of the sibs of our patient had normal colour vision, we consider the coincidental occurrence of hereditary colour-blindness unlikely, even though we were unable to examine all the members of the family. In a large pedigree with Friedrich's ataxia, Heck (1963) reported the simultaneous occurrence of retinal degeneration and blue-yellow colour-blindness, which was transmitted independently of the ataxia, through an intermediate X-linked mechanism. The heterozygous females exhibited either pigmentary degeneration or tritania. The normal eye findings in the mother of our patient speaks against this mode of transmission.

Blue-yellow colour-blindness is known to occur in advanced cases of pigmentary degenerations of the retina (Duke-Elder, 1967). Even though the ophthalmoscopic findings in our patient were not very severe, degenerative changes in the retinal cones appear to be the most likely explanation of the colour-blindness. In a condition where there is generalized metabolic disturbance of the retinal photoreceptors, it is reasonable to assume that the cones and the rods can be equally affected. If this assumption is correct the absence of the description...
of a similar finding in other reported cases of a-\(\beta\)-lipoproteinæmia remains unexplained.

**Summary**

A 15-year-old Arab boy born to a first-cousin marriage and suffering from a-\(\beta\)-lipoproteinæmia is described. In addition to stethorræha, ataxia, skeletal deformities, and night blindness he had defective blue-yellow colour-vision, a finding not described in previous reports on a-\(\beta\)-lipoproteinæmia.

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**Copper Poppers: A Benign Cause of Blue Diapers**

The blue diaper syndrome has been described in one family by Drummond *et al.* (1964). The syndrome was apparently caused by a gastro-intestinal transport defect in the absorption of tryptophan so that unabsorbed tryptophan in the gut was converted to indoles by intestinal bacteria.

The indoles were then absorbed and converted in the liver into indican which was then excreted in the urine. Conjugation of two molecules of indican in the urine produced the water insoluble dye indigotin, which was responsible for the blue discoloration of the diapers.

This note describes a new cause for blue diapers which is not due to an inborn error of metabolism, but to a chemical interaction, under certain circumstances, between normal urine and the metal snaps which are sometimes used for securing infant nappies.

This condition was brought to our attention when a female infant was seen for a routine post-natal clinic checkup at the age of 3 months. At that time, the patient appeared to be perfectly healthy, but the mother complained that the nappies were sometimes turning blue after the child had urinated. The child was admitted to hospital and a number of investigations, including tryptophan tolerance tests, were undertaken. These were all normal. At that time, the mother declared that the blue coloration was particularly obvious around the metal poppers which were used for securing the napkin. This observation led us to postulate that a chemical interaction was occurring between a constituent of the urine and the metal in the poppers. This led to a number of investigations.

**Results**

1. Following an oral L-tryptophan tolerance test (100 mg/kg) the urine was tested for indican by Jaffe’s method (Hawk and Bergeim, 1931). No excess indican was present and the tryptophan metabolite pattern was normal on paper chromatography.

2. Examination of a napkin from the baby showed that the colour occurred in patches which were most marked around the metal poppers. Attempts were made to elute the colour from a blue stained napkin with acetone, benzene, and chloroform. The blue material was insoluble in these solvents but was readily soluble in 1 N ammonium hydroxide. On the addition of 1 N hydrochloric acid to the solution, the blue colour disappeared but returned again when it was realkalinized with sodium hydroxide. The colour also disappeared on the addition of aqueous sodium cyanide (Curtman, 1938).

3. Dry, powdered di-ethyl di-thio carbamate was added to the blue eluate from a stained napkin. A yellow colour was produced indicating the presence of copper in the solution.

4. A cleaned popper from the child’s napkin was soaked in 1 N ammonium hydroxide and a strong