the baby cries with the angle of mouth being pulled down to the sound side due to unopposed action of depresor anguli oris muscle(2,4).

The importance of identifying this anomaly is that it is associated with other congenital malformations in over 20% of cases, most commonly being associated with cardiovascular anomalies and congenital dislocation of hip. Of the 44 infants with this syndrome, Pape and Pickering found 27 to have major anomaly of skeletal, genitourinary, respiratory and cardiovascular systems. The disorder most commonly associated with this facial defect is congenital heart disease, the commonest defect being ventricular septal defect(2,4).

In this baby, depressor anguli oris muscle deficiency was associated with congenital heart disease in the form of truncus arteriosus and other anomalies such as polydactyly, suggesting an embryonic defect affecting multiple organ system. To our knowledge the combination of defects which we have described in this baby has not been reported in literature.

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Alkaptonuria: Early Detection

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Alkaptonuria is an inborn error of tyrosine metabolism resulting from deficiency of the enzyme homogentisic acid oxidase which is necessary for converting homogentistic acid into malylaceto-acetic acid(l,3).

Case Report

A one-month-old boy born to non-consanguinous normal parents, was brought with the complaint of slight alteration in urine color. He was the product of full term normal delivery. His weight, length, armspan and other anthropometric measurements, as well as general and systemic examination revealed no abnormality.

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The urine was straw colored when voided and turned dark brown to black on standing for 5-6 hours. The baby was, therefore, suspected to suffer from alkaptonuria. To confirm the diagnosis the following investigations were performed on freshly voided urine samples: (i) Addition of alkali turned the color of urine dark black within 10 min; (ii) Filter paper impregnated with 10% sodium hydroxide turned black within 5 minutes when dipped into urine; (iii) Benedict's test was strongly positive with red brown precipitate at bottom and black colored supernatent; (iv) Glucose oxidase test (with multistix) was negative; (v) Fehling's test was positive; (vi) Addition of dilute ferric chloride solution drop by drop showed an evanescent violet blue color; (vii) Addition of equal volume of ammoniacal silver nitrate quickly produced black precipitate; *(viii) A* sensitized photographic film turned to black when urine dropped on it; and *(ix)* Paper chromatography of urine demonstrated presence of homogentisic acid and 2:4-dihydroxy phenyl pyruvic acid. Similar tests performed on parents and other family members (elder brother and sister of patient) were negative.

Discussion

Alkaptonuria is usually an autosomal recessive disorder though in a minority of cases it is transmitted as autosomal dominant(l,4). The aminoacids tyrosine and phenylalanine are not metabolized beyond the stage of homogentisic acid which is, therefore, excreted in urine. Homogentisic acid is a strong reducing agent, which on exposure to atmospheric oxygen for some hours, gets converted to an oxidized polymer that is black in color. Urine containing homogentisic acid, therefore, turns black on standing. Levels of homogentisic acid in blood are minimally increased because it is rapidly cleared by the kidneys.

Alkaptonuria is suspected clinically in adults if ochronosis: dark colored spots on the sclera and diffuse dark pigmentation of conjunctiva, ear cartilage and nose are seen. This pigmentation occurs because of binding of homogentisic acid and its oxidized polymer to collagen. Complications are early calcification of cardiac valves with chronic mitral and aortic valvulitis, early generalized arteriosclerosis, renal stones, chronic prostatitis and nephrosis. Alkaptonuria has been reported in association with hyperuricemia, polycythemia, Addison's disease, diabetes mellitus and ankylosing spondylitis in adults(4).

So far no treatment is available to treat the condition. Since ascorbic acid impedes oxidation and polymerization of homogentistic acid *in vitro,* its use has been suggested as a possible means of decreasing pigment formation and deposition(l,5), but its efficacy has not been established(l). Corticosteroids have been used to prevent the disabling complications of this disorder.

Awareness and early detection help in allaying parenteral anxiety. Administration of ascorbic acid right in the neonatal period may help in preventing complications of this progressive disabling metabolic disorder at later ages.

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Efficacy of Halofantrine in Malaria

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Malaria continues to be a major health problem in the tropical countries. In India, about 2 million suffer from the disease every year; the reported cases in 1989 being 20,17,823 with 268 deaths and in 1990, 17,77,263 cases and 222 deaths(l). Thirty five per cent of the total cases of malaria in our country occur in children below 15 years of age(2). Of late, more and more cases appear to be resistant to chloroquine and also to other antimalarials(3). This is particularly so with *Plasmodium falciparum* malaria(4). In India, chloroquine resistant strains of *P. falciparum* were demonstrated in 19 states of the country in 1986(5). Halofantrine, a phenanthrene-methanol, is an orally administered schizonticidal drug, effective against both chloroquine sensitive and resistant strains of *Plasmodia.* We report our results with 46 children suffering from malaria treated with halofantrine using a 3 dose regimen of 8 mg/kg 6 hourly(6,7).

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Material and Methods

Forty six children suffering from malaria caused by *Plasmodium vivax* and/or *Plasmodium falciparum* or *Plasmodium ovale* were included. The children attended the Outpatient Department or were admitted to the Pediatric ward of Sassoon General Hospitals, Pune. The criteria used for selection were history of fever and the presence of malarial parasite on peripheral smear (gametocytes or asexual forms).