

sions was described in a 4-month-old(8). Mohanty has studied a large series, the youngest being a boy of 3 years(9).

Multiple osteolytic lesions mimic a variety of childhood malignant conditions like neuroblastoma, histiocytosis, lymphoreticular malignancies, syphilitic, pyogenic or mycotic osteomyelitis. Though the tubercular lesions are disseminated, the response to therapy is excellent and prognosis is good as was seen in our case.

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## Cohen Syndrome

**Said Alavi**

**A. Kher**

**A. Kumar**

**M. Muranjan**

**B. Bharucha**

In 1973, Cohen *et al.*(1) described three cases of a syndrome, the major features of which were obesity, hypotonia and mental retardation with facial, oral, ocular and limb anomalies. Eighty cases have since been reported(2) and inheritance is autosomal recessive(3).

This report profiles two cases that have most of the major abnormalities and is the first report from India after the syndrome has been recognised.

### Case Reports

**Case 1:** A ten-year-old male child of non-consanguineous parentage was referred

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*From the Departments of Pediatrics, K.E.M. Hospital and Seth G.S. Medical College, Parel, Bombay 400 012.*

*Reprint requests: Dr. B.A. Bharucha, Associate Professor, Department of Pediatrics, K.E.M. Hospital, Parel, Bombay 400 012.*

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to the Genetic Clinic with history of defective vision since early infancy and delay in attaining milestones. His four siblings and parents were normal. He had been delivered by cesarian section for prolonged labour and had a birth weight of 4 kg. There was apparently no birth asphyxia or neonatal morbidity. Motor, intellectual and language milestones were delayed. Dys-

morphic abnormalities are shown in the *Table*. IQ (using Kamat's test) was 35-40, optic fundi showed bilateral pigmentary degeneration with optic atrophy. Barr body was negative. Skeletal X-rays, ultrasonography of abdomen, 2-D echocardiogram and plasma and urine aminacido-grams were normal.

**Case 2 :** An eight year old female child of non-consanguinous parentage was referred to the Genetic Clinic with scholastic backwardness and delayed milestones without family history of similar complaints. The mother's antenatal history was normal. The patient was delivered in hospital at term with birth weight of 2.25 kg without any complications. There was delayed achievement of milestones. Major physical findings are shown in the *Table*. Investigations revealed normal optic fundi and audiometry, social maturity by Vineland social maturity scale of 46-48 and normal abdominal ultrasound, 2-D echocardiography, skeletal X-rays and urine and plasma aminoacidogram. Karyotype from peripheral blood leucocytes was 46 XX.

### Discussion

Both the patients described in this report show most of the features described by Cohen *et al.*(1) with obesity, mental retardation and limb and craniofacial abnormalities(4). The three most striking features that help in the recognition of the syndrome in a mentally retarded child are hypotonia, obesity and prominent central incisors (*Fig.*).

Other commonly encountered syndromes with obesity, mental retardation and hypogenitalism that have to be considered in the differential diagnosis are—the Prader-Willi syndrome, which was ruled out because of the absence of small hands and feet and characteristic craniofacial

TABLE—Summary of Cases

Major features in Cohen syndrome	Case 1	Case 2	Cohen <i>et al.</i> (1) 1973
<b>Growth and Performance</b>			
Obesity	+	+	3/3
Short stature	-	-	1/3
Mental retardation	+	+	3/3
Delayed puberty	?	?	2/3
<b>Orofacial</b>			
Microcephaly	+	+	3/3
High nasal bridge	+	+	3/3
Maxillary/malar hypoplasia	+	+	3/3
Short philtrum	-	+	3/3
Micrognathia	+	+	3/3
Open mouth	+	+	3/3
Prominent central incisor teeth	+	+	3/3
Narrow palate	+	+	3/3
<b>Ocular</b>			
Downslanting eyes	-	-	3/3
Strabismus	+	-	2/3
Pigmentary degeneration	+	-	2/3
Myopia	-	-	1/3
Optic atrophy	+	-	2/3
<b>Limbs and skeleton</b>			
Hypotonia	+	+	3/3
Joint laxity	+	-	3/3
Cubitus valgus	+	+	3/3
Genu valgum	+	+	3/3
Narrow hands and feet	+	+	3/3
Syndactyly	-	-	1/3
Scoliosis	-	-	3/3



*Fig. Prominent large central incisor teeth in Cohen Syndrome.*

features that are essential for the diagnosis(5), the Börjeson-Forssman-Lehmann syndrome, which was ruled out due to the absence of coarse facies, large ears and soft tapering fingers(6) and the Bardet-Biedl syndrome, which again was ruled out because of the absence of post-axial polydactyly. Progressive retinitis pigmentosa is seen in Bardet-Biedl syndrome while in Cohen syndrome, there is chorioretinal dystrophy with pigmentary degeneration and optic atrophy(7).

The prominent central incisors in the wide open mouth seems to be the most striking feature for identification of the individual. De novo reciprocal 5q-7p translocation has been reported recently in a fifteen-year-old girl with Cohen syndrome(8).

The mental handicap in Cohen syndrome can be of severe degree and weakness and hypotonia persist beyond infancy.

The discovery of a visual problem poses a further handicap. No other major sequelae have been noted so far. Precise diagnosis on the basis of simple identifiable clinical features favors accurate genetic counselling and offers relatively benign prognosis.

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## Knowledge of Community Based Practitioners Regarding Vaccine Preventable Diseases

Harmesh Singh  
P.J.S. Gill

Diphtheria, pertussis, tetanus, poliomyelitis, measles and tuberculosis continue to kill or cripple millions in the developing countries(1). These are preventable infections because safe, effective and easily administrable vaccines are available. Starfield(2) has reported that nearly half of

the children are seen by a generalist physician regularly, whereas fewer than 1/3 are cared for by a pediatrician and 10% receive care related to place rather than a particular physician. In our country also it may be true that many children receive care related to place rather than a particular physician probably due to a long standing relationship of trust, shortage of time as well as resources, illiteracy and ignorance. The knowledge of these community based primary practitioners has an important impact on prevention and control of the six vaccine preventable diseases. In order to evaluate the knowledge of these practitioners, about vaccine preventable diseases, we planned this study.

### Material and Methods

Forty six community based primary practitioners doing general practice from various localities of Ludhiana were selected randomly. Clinic to clinic visits were carried out by the authors. During these visits the practitioners were interviewed in detail regarding symptoms, preventive measures and treatment of the six killer diseases and schedule, contraindication and storage of various vaccines. The proof of their qualifications and experience could often not be documented.

### Results

All the practitioners interviewed were males. Their qualifications included Registered Medical Practitioners in 23.9%, Bachelor of Ayurved Medical Science

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*From the Departments of Pediatrics and Family Welfare and Postpartum Program, Dayanand Medical College and Hospital, Ludhiana.*

*Reprint requests: Dr. Harmesh Singh, C/o Ganesh Medical Hall, Tagore Nagar Extension I, Ludhiana (Punjab).*

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