this decision was provided to the parents throughout many visits. Patients with Down’s syndrome may have cryptorchidism or a small penis but other genital developmental abnormalities are uncommon (Hamerton, 1971). Only one case of pseudohermaphroditism in a patient with Down’s syndrome has been reported previously and that was a female with congenital adrenal hyperplasia (Srivuthana, et al, 1971). There is some evidence, however, that the association of sex chromosome cryptorchidism lack of reported cases with the association of her mother became pregnant by her mechanism their two abnormal states seen in our patient suggests et al, 1971). There is some evidence, however, that congenital risk of bearing another child with increased amniocentesis was performed because of her in- affected if the mother was the syndrome was 46,XY. The possibility was 46,XY. The possibility of bearing another child with aneuploidy and cells was 46,XY. The possibility of bearing another child with Down’s syndrome reported we feel that if these abnormalities existed more abnormalities if the mother was the carrier of an X-linked form of male pseudohermaphroditism was discussed with the parents. A normal male infant was delivered at term.

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Möbius Syndrome with Poland’s Anomaly*

Summary. A five-year-old boy with Möbius syndrome, Poland’s anomaly, and dextrocardia is described. These malformations have not been previously reported. The propositus had ipsilateral absence of the sternal portion of the pectoralis major muscle associated with acrominia, syndactyly, brachydactyly of the index, middle, ring, and fifth finger, as well as radiological evidence of hypoplasia of the index middle and ring fingers, associated with syndactyilia and absent middle phalanges.

Möbius syndrome associated with Poland’s anomaly, as represented by the case reported here, has not been previously described.

The first report of congenital bilateral facial paralysis was made by von Graefe in 1880. In 1888, Möbius emphasized the nuclear agenesia of the cranial nerves. Some of the most comprehensive reviews of this rare disorder were made by Henderson in 1939, Danis in 1945, Gorlin and Pindborg in 1964 as well as others (Hellström, 1949; Richards, 1953; Evans, 1955).

Alfred Poland in 1841, dissected the body of a criminal with unilateral symbrachydactyly associated with ipsilateral (same side) aplasia of the sternal head of the pectoralis major muscle. In 1900, Fürst provided a detailed analysis of the type of hand malformation, the associated muscle defects and insight into the aetiology.

Case Report

This 5-year-old boy was first seen at the Orthopaedic Hospital, Los Angeles, California in May 1971, because of multiple congenital abnormalities involving the eye, facial muscles, jaw, teeth, palate, sternum, chest, and hand. The mother’s pregnancy was apparently normal. She gained approximately 15-9 kg in weight from an initial weight of 66.7 kg. The father and mother were unrelated, and were both 30 years of age at the time of the patient’s birth. The birthweight was 40-0 kg. The baby breathed and cried spontaneously. No diagnosis of any syndrome was made. He had two operations on his eye muscles for stabismus, at age 1 and 3 years. There was no family history of neurological or somatic disorders.

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Physical Examination. The 5-year-old boy was alert, co-operative, normally intelligent, but somewhat sensitive about his abnormalities. He had the following congenital abnormalities: (1) fixed, expressionless facies (Fig. 1a) with inability of the eyes to move on lateral or medial gaze; (2) inability to close the lids completely (Fig. 1b); (3) bilateral facial nerve paralysis with inability to smile; (4) microlingua; (5) hypoplasia of the teeth; (6) high arched palate; (7) micrognathia and hypoplastic mandibles; (8) obesity; (9) short, fixed sternum with aplasia of the sternal portion of the left pectoralis major muscle (Figs. 2a and 2b); (10) dextrocardia without murmurs; (11) left-hand acromicria with soft tissue syndactyly and brachydactyly of the second, third, fourth, and fifth fingers (Fig. 3). He also had slight clubbing of the left fingernails and (12) involvement of the cranial nerves III, IV, V, VI, VII, IX, and XII.

His vision, hearing, and sense of smell were intact. He...
weighed 23.4 kg was 106.6 cm tall with a head circumference of 53.3 cm and a chest circumference of 60.3 cm. The sternocleidomastoid and trapezius muscles were normal. He had no physical signs of situs inversus or congenital heart disorder. The genitalia were normal.

**Radiographic Study (2 October 1972)**

**Chest: PA and Lateral Views.** The heart was prominent in the right chest and was rotated to the right (Fig. 4). The aortic arch appeared on the left. Soft tissues of the left chest were much thinner than those on the right. Pulmonary vasculature was within normal limits and the lungs were clear. The right ribs laterally were a little closer together than the left. There was mild scoliosis. The liver was identifiable on the right with stomach gas bubble present on the left. The sternum was quite short and the segments were fused, including the manubrium. There was anterior convexity of the sternum just below the junction with manubrium.

**Skull Series.** Cranial vault was of average size. The sutures were open and were not spread. The sella turica had normal appearance. There were no abnormal intracranial calcifications. The mandibular angles were quite obtuse.

**Left hand: AP, Lateral, and Oblique Views.** Anatomy of the carpal ossification centres in the distal radius and ulna was unremarkable. There were five metacarpals with finger metacarpals approximately the same length. There was less than the usual amount of modelling of the index and middle metacarpals. An accessory ossification centre was present at the base of the second metacarpal and at the distal end of the first metacarpal which also showed lack of normal modelling. The thumb had two phalanges, the proximal thumb phalanx showed an accessory ossification at the distal end. The index, middle, and ring fingers had only two phalanges, the proximal phalanges being slender and the other phalanges quite hypoplastic (Fig. 5). There was syndactyly of these three fingers and together they present a short projecting stump in the middle of the hand. The little finger had three phalanges, although the space between the middle phalanx and the distal phalanx was quite small and may eventually coalesce.

**Fig. 4.** Radiograph of the chest revealing dextrocardia and decreased soft tissue of the left side of chest consistent with absence of muscle mass; also short fused sternum.

**Fig. 5.** Radiograph of the left hand. Note phalangeal absence of the distal second to fourth rays and interphalangeal webbing with hypoplasia most marked in the index, middle, and ring fingers with syndactyly, missing middle phalanges and variations of modelling.
Case Reports

Discussion

Our patient had typical features of Möbius syndrome (Table I) as well as Poland's anomaly (Table II), an unusual association and heretofore unreported. In addition, dextrocardia has not been previously reported in either Möbius syndrome or Poland's anomaly. Möbius syndrome has been reported with arthrogryposis multiplex congenita, Klippel-Feil syndrome (Evans, 1955) and fascioscapulohumeral muscular dystrophy (Hanson and Rowland, 1971).

TABLE I

MÖBIUS SYNDROME

| Congenital facial palsy (Craniat nerve VII) |
| Difficulty in swallowing (Craniat nerves IX and X) |
| External ophthalmoplegia (Craniat nerves VI) |
| Other cranial nerves VII, III, and V |

Associated abnormalities

1. Clubfoot
2. Epicanthal folds
3. Hypertelorism
4. Strabismus
5. Ear abnormalities
6. Micrognathia
7. Bilid uvula
8. Palaral palse
9. Perodactyly, syndactyly, brachydactyly
10. Congenital heart disease
11. Pectoral muscle hypoplasia

TABLE II

POLAND'S ANOMALY AND ASSOCIATED FINDINGS

I. Muscle
a. Absence or hypoplasia of pectoralis major (sternal head)
b. Absence or hypoplasia of pectoralis minor
c. Absence or hypoplasia of external abdominal, latissimus dorsi, serratus anterior, intercostal

II. Skeletal
a. Syndactyly—brachydactyly
b. Absence or hypoplasia of rib
c. Hemivertebra
d. Absence or hypoplasia of radius, ulna, carpal, metacarpal, phalange
e. Finger webbing
f. Sprengel's deformity

III. Skin
a. Café-au-lait pigmentation
b. Absence of nipples
c. Webbing of axilla
d. Absence of subcutaneous tissue

IV. Genitourinary
a. Renal aplasia or hypoplasia (Temtamy and McKusick, 1969; Mace et al, 1972)
b. Pyelonephritis (Temtamy and McKusick, 1969)
c. Undescended testes

V. Others
a. Herniation of lung (Ehrenhaft et al, 1966)
b. Acute lymphoblastic leukemia (Mace et al, 1972)
c. Absence of the breast or asymmetry
d. Hemifacial atrophy
e. Dextrocardia (not previously reported)

Jorgenson (1971) reported a patient with Möbius syndrome associated with bilateral hand defects and hypoplasia of the right pectoral muscles. All the fingers of the right hand were missing at the metacarpophalangeal joints except for the thumb which was slightly hypoplastic (ectrodactyly). The left hand had a normal thumb and forefinger but the other three digits were hypoplastic with interdigital webbing. In contrast, our patient with Möbius syndrome had ipsilateral absence of the sternal portion of the pectoralis major muscle associated with acromia, syndactyly, brachydactyly of the index, middle, ring, and fifth digits as well as radiological evidence of hypoplasia of the left index, middle, and ring fingers with syndactyly, absent middle phalanges, and variations of modelling.

Additional associated malformations occurring with Möbius and Poland's anomaly have been summarized in Table I and Table II. Various isolated sporadic malformations have also been reported: unilateral symbrachydactyly, bilateral aplasia of the sternal head of the pectoralis major muscle, and bilateral ectrodactyly with a unilateral pectoral muscle defect (Ehrenhaft, Rossi, and Lawrence, 1966).

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Familial Bird-headed Dwarfism
(Seckel's Syndrome)

Summary. Low birth weight dwarfism with mental retardation, large eyes, a beaklike nose, narrow face, receding mandible, and dental anomalies are the specific features of 'bird-headed dwarf of Seckel'. The following case report presents details of a Seckel dwarf with familial occurrence of the trait and, thus supports an autosomal recessive mode of inheritance. In addition, the possible significance of dental alterations is noted.

Case Report

The proposita is a 46-month-old negro who possessed the clinical syndrome of Seckel's bird-headed dwarfism; the familial occurrence of the trait in three sibs is also included.

The patient was born 2 May 1969 in Osceola, Arkansas. The child weighed 1308 g at birth and 40 weeks' gestation. To date the patient presented as severely retarded, and weighing 4578 g.

The proposita had microcephaly and oxycephaly, with pro-ophtosis of the eyes, nystagmus, and lobeless ears (Fig. 1). Marked micrognathia was noted as well as a central posterior cleft of the palate (Fig. 2). The dentition revealed severe hypoplasia of enamel in the primary dentition; however primary second molars showed no

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