INTRODUCTION
Poland syndrome consists of congenital unilateral absence or hypoplasia of the sternal head of the pectoralis major muscle and, in the majority of patients, a variable degree of ipsilateral hand and digit anomalies.1 Mobius syndrome is a rare disorder characterized by congenital non-progressive unilateral or bilateral VI and VII cranial nerve paresis with or without involvement of other cranial nerves.2 The estimated prevalence of Poland-Mobius syndrome is 1: 500,000.1 The most obvious symptom is loss of facial expression; the ocular symptoms include restricted bilateral eye movements, ptosis, nystagmus, and inability to close the eyelids. Mild to moderate mental retardation occurs in 10% of the cases.2 Intelligence may be normal and most people lead full lives with professional and personal success.3

This rare syndrome is hereby described in a newborn.

CASE REPORT
A two days old male baby was admitted via outpatient department with difficulty in feeding, mouth deviation and inability to close eyes.

He was born to a 29 years old mother at the gestational age of 38 weeks by spontaneous vaginal delivery at hospital. There was no history of birth asphyxia. The pregnancy was uncomplicated and there was no history of any drugs, smoking/teratogens intake during pregnancy. His parents were not related and healthy. He had one elder healthy sister. There was no history of any dysmorphism in family.

On presentation, growth parameters were at 25th centiles; (weight: 3.1 kg, length: 51 cm, HC: 34 cm). Examination revealed several congenital abnormalities. He had an expressionless face, deviation of mouth towards right side and high arched palate (Figure 1). Neither eye could be abducted beyond the mid line (Figure 2). The right chest wall had a defect in the muscular layer and hand was absent of the right upper limb (hand acheiria, Figure 3). Rest of his examination including other cranial nerves and limbs was normal. Cornea were clear. He had no visceromegaly; the cardiovascular examination and genitals were normal.

MRI brain, echocardiography and abdominal ultrasound scan were also normal. Chromosomal analysis showed normal XY karyotype. He was diagnosed as a case of Poland-Mobius syndrome on the basis of facial nerve and bilateral VI nerve involvement along with associated limb and chest deformities.

DISCUSSION
Mobius syndrome was first defined by Paul Julius Mobius, a German neurologist, in 1888, who reported patients with congenital, non-progressive, bilateral VII and VI nerve palsy. Primary criteria for diagnosing Mobius syndrome are facial palsy with impairment of ocular abduction. Dysfunction of other cranial nerves and orofacial abnormalities are commonly associated but not necessary for the diagnosis.4

Abramson et al. classified and graded the syndrome on the basis of clinical findings of cranial nerves and musculoskeletal anomalies using the acronym CLUFT (Cranial nerves, Lower limb, Upper limb, Face and Thorax).5

Etiology of Mobius syndrome is multifactorial with several proposed theories. The most supported one is transient ischemia or hypoxic insult to fetus. The use of drugs may also be linked to the development of Mobius syndrome. Use of misoprostol or thalidomide by women during pregnancy increases incidence dramatically. Incidence of Mobius syndrome is 1: 50,000 to 100,000.6

Most cases of Mobius syndrome are sporadic but familial cases are also reported with autosomal dominant, auto-
recessive and x-linked recessive modes of inheritances. In addition to the involvement of chromosome 13, other loci map to 3q21-q22 and 10q21.3-q22.1.2

Clinical features of Mobius syndrome includes complete or partial unilateral or bilateral facial nerve palsy with bilateral VI nerve palsy with or without involvement of other cranial nerves; III, IV, V, VIII, IX and X and physical abnormalities like multiple limb malformations including syndactyly, absent digits, and talipes.2 Chewing, swallowing, and coughing difficulties may lead to respiratory compromise later in life. They also have hearing and speech problems. Mental retardation, autism and behavioral problems are also seen.2

Poland syndrome was first described in 1841 by Alfred Poland who dissected the body of a criminal with unilateral symbrachydactyly associated with ipsilateral aplasia of the sternal head of the pectoralis muscle. Other anomalies associated with this syndrome include hypoplasia of the hand and forearm, hypoplasia of the breast, bilateral epicantus, equinovarus, Mobius syndrome, upper musculature anomalies, rib cage deformities, and an absence of the ipsilateral kidney.6 MRI findings may demonstrate hypoplasia of pons particularly in its dorsal aspect where the region of the facial colliculus and 6th nerve complexes.7

McGilivary and Lowary defined the incidence of Poland syndrome of 1 per 32,000 live births.8 Boys are affected more than girls. The syndactyly is usually in the right hand as in this patient. The inheritance of Poland syndrome is entirely sporadic.1 Bavnik and Weaver suggested that Poland, Klippel-feil, and Mobius sequences should be grouped together on the basis of similar developmental pathogenesis referred to as the subclavian artery disruption sequence.9

Treatment is conservative consisting of reconstruction and plastic surgery of the breast, hands and fingers for cosmetic purposes. If the infant has feeding difficulties, physical and occupational therapy is required. Surgery can correct ophthalmological problems and smile surgery can be done which includes muscle transfer from the thigh to the corner of the mouth. Surgery is ideally performed in patients just before they reach school age at 4-5 years.10

REFERENCES

Figure 1: Bilateral VI nerve palsy, resulting in complete loss of abduction.
Figure 2: Picture showing left facial palsy.
Figure 3: Chest deformity and abnormalities of the right hand.