Möbius syndrome or congenital facial paralysis is a rare deformity. The exact frequency of occurrence is not known, with only several hundred cases being reported in the literature. This type of facial paralysis has been classically described as bilateral, complete, and permanent. However, the paralysis occasionally may be asymmetric, in which the upper face is affected and the lower lip movement spared. Although uncommon, this syndrome is very distinct.

Bilateral congenital facial paralysis has traditionally been associated with Paul Julius Möbius. In 1888, Möbius described a clinical entity involving bilateral paralysis of the sixth and seventh cranial nerves. He was the first to single out these particular palsies as a separate entity. Hence, the hallmark clinical features of Möbius syndrome are paralysis of the sixth and seventh cranial nerves. The seventh cranial nerve or facial nerve innervates the muscles of facial expression. Paralysis of this nerve results in the mask-like facies and the inability to smile. The paralysis of the sixth cranial nerve results in the inability to abduct the eyes or simply to look to the side by just moving the eyes. Because of this inability to move the eyes laterally, the child follows objects by turning his head from side to side. Patients with Möbius syndrome may also have additional problems. The paralysis may involve other cranial nerves such as the nerve that innervates the tongue. Limb anomalies have been reported in 25% of cases with the most common problem being the club foot. Additional problems can be lack of development of chest musculature such as the pectoral muscles which was present in 15% of the reported cases.

The etiology of this syndrome remains unclear. One hypothesis is that nuclear hypoplasia or agenesis of the sixth and seventh facial nerves causes the anomaly. Another hypothesis is primary aplasia or lack of development of the muscles leading to atrophy of the nerve. Also in doubt is the role that heredity plays. While many feel heredity plays no role, there have been reports of a small number of cases occurring in families indicating a possible hereditary tendency.

The diagnosis of Möbius syndrome is not always made in the newborn nursery and is often not recognized as Möbius syndrome for several months. The parents usually show the most concern over the mask-like facies which is due to the paralysis of the facial nerve. The sixth nerve paralysis becomes obvious when the child follows objects by turning his head from side to side. The child may experience eating problems when food becomes lodged in the cheeks requiring manual removal. However, in the congenital form of facial paralysis, functional problems are often not as significant as might be expected. Corneal protection which is related to the ability to close the eyes is usually adequate with problems related to corneal exposure uncommon. It is the lack of facial expression or inability to smile that poses the major problem. This can severely inhibit psychosocial development as the child approaches school age and thus becomes aware that they are different. We feel this obstacle to interpersonal relationships and social development should play a role in the timing of surgical intervention.

The timing of recommended surgical correction is of course influenced by the age of the patient at referral. Ideally, one would want to evaluate Möbius syndrome patients as infants. Due to the psychosocial problems that can result from bilateral facial paralysis, we recommend early surgical intervention if possible. It
is preferable to have the child’s reconstruction complete before he starts school. This would mean starting the reconstruction at four to five years of age. However, the age of the patient does not effect the operative outcome; successful reconstruction can be performed in adults as well.

The treatment of patients with Möbius syndrome is directed toward restoration of facial motion. Acquired facial paralysis differs from the congenital type found in Möbius syndrome, and its treatment will not be discussed in this article. In the congenital cases, corneal exposure is seldom a problem and therefore does not require surgical treatment. However, in acquired cases eye problems are frequent and generally require surgical intervention. In bilateral congenital facial paralysis the major goal is to restore motion in the appropriate direction to the upper lip and commissure. In unilateral cases, a nerve graft from the functioning facial nerve to the effected side can be performed. However, in bilateral cases such as Möbius syndrome, there is no functioning facial nerve available. The surgical options in these cases are a local muscle transfer procedure or free muscle transplantation. Local regional muscles such as the temporalis or masseter can be used to restore facial movement. Either of these muscles of mastication can be moved maintaining their innervation to restore movement of the upper lip and commissure. Retraining and practice is necessary to get the maximum benefit.

A newer procedure is microvascular muscle transplantation. This involves moving a muscle from another part of the body along with its nerve and blood supply. The nerve and vessels to the muscle are reconnected with the aid of a microscope. The muscle is oriented in such a fashion to provide motion to the upper lip and commissure.

In each of these procedures the family needs to have realistic expectations of what can be accomplished. If successful, these procedures can provide tone to the upper lip and commissure at rest, with muscle action that can produce a smile. Fine movements or variation in expression are usually not possible. However, these procedures can make dramatic improvements in the mask-like facies of Möbius syndrome. They provide the potential benefits of facial animation with improved social interaction.