

소아재활

게시일시 및 장소 : 10 월 18 일(금) 13:15-18:00 Room G(3F)

질의응답 일시 및 장소 : 10 월 18 일(금) 16:13-16:17 Room G(3F)

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Rehabilitation of an Infant with Poland-Moebius Syndrome: A Case Report

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Objective

Poland-Moebius syndrome is a rare congenital disease showing both features of Poland syndrome and Moebius syndrome. Poland syndrome displays absence of pectoralis major muscle, hand anomalies, and malformation of internal organs. Moebius syndrome, described as the degeneration of nuclei of facial nerve and abducent nerve, typically demonstrates bilateral nonprogressive facial palsy and loss of abduction of the eye. Poland-Moebius syndrome has been estimated to occur in 1 out of 500,000 persons. Due to its rareness, neither the course of the disease nor well-established treatment plans is known. We aimed to report catch-up milestone of a child with Poland-Moebius syndrome after 15 months of comprehensive and individualized rehabilitation management.

Case description

A 2-month-old boy with Poland-Moebius syndrome was referred to our hospital for rehabilitation. He featured bilateral facial palsy, both esotropia (Fig. 1), left club foot (Fig. 2), right hand hypoplasia with syndactyly (Fig. 3), and marked left hydronephrosis. He was clearly conscious and the movement of all four limbs was active without muscle weakness. Hyperreflexia or spasticity was not checked. On Denver developmental screening test, gross motor was measured 1 month, fine motor 2 months, social 1 month, and language 1 month. We did further evaluations to find accompanied deformities. On videofluoroscopic swallowing study, penetration or aspiration was not observed. Nerve conduction study revealed incomplete both facial and trigeminal neuropathies. There was no leg length discrepancy on serial scannograms. Comprehensive rehabilitation was performed for 15 months. Physical therapy consisted of head control training, stretching and strengthening exercises, and mobility exercise. Occupational therapy included facial massage, oromotor facilitation, fine motor training, and various sensory stimulation. For the club foot, he underwent percutaneous achilles tendon lengthening surgery at the age of 2 months and had worn Denis-brown orthosis for 14 months. After the rehabilitation, gross motor was assessed 9 months, fine motor 16 months, social 18 months, and language 12 months at birth age of 16 months. He took off the foot orthosis. An operation for syndactyly is planned for him.

Conclusion

We report comprehensive and individualized rehabilitation management of an infant with Poland-Moebius syndrome. Despite several problems affecting normal development, overall developmental assessment showed catch-up milestone. Considering the diversity of symptoms of Poland-Moebius syndrome, specialized diagnostic tests and individualized rehabilitation directed toward the specific abnormalities by multidisciplinary team approach should be needed.

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Figure 1. Bilateral facial palsy and both esotropia are shown in the face of the boy with Poland-Moebius syndrome.



Figure 2. Left club foot deformity is seen.



Figure 3. Right hand hypoplasia with syndactyly is displayed.