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Mouthstick prosthesis placement in a 19-month-old arthrogryposis multiplex congenita patient: case report

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Abstract

Arthrogryposis multiplex congenita is a congenital disorder distinguished by generalized joint immobility and skeletal muscle hypoplasia. Although the afflicted individual has normal intelligence, physical constraints often prevent independent living. This case report describes the successful fabrication and placement of a mouthstick prosthesis in a 19-month-old patient.

Arthrogryposis multiplex congenita (AMC) is a congenital disorder distinguished by limited joint mobility and a generalized hypoplasia of skeletal muscles. The term arthrogryposis is derived from the Greek word meaning bent or curved joint. It was first described in 1841 as a congenital myodystrophia (Otto 1847). This rare disorder, occurring in 3 of every 10,000 births, often results in multiple joint deformities involving the hands, wrists, elbows, hips, knees, and feet (Laiten and Himensalo 1966). Clubbing of the hand and foot are also frequent occurrences of the disorder. Associated causes of AMC are:

1. Neurologic abnormality such as a meningomyelocele, anterior motor horn cell deficiency, prenatal spasticity, or gross brain defect
2. Muscular problems such as muscle agenesis, fetal myopathies, or myotonic dystrophy
3. Joint and contiguous tissue problems, such as synostosis, lack of joint development, and aberrant fixation of the joints
4. Fetal crowding and constraint such as with multiple births or with oligohydramnios in disorders such as renal agenesis or early persistent leakage of amniotic fluid (Thomas and Smith 1974; Pena and Shokeir 1976; Edmonson and Crenshaw 1977; Brown et al. 1980; Williams and Holmes 1980).

Treatment of the AMC patient involves the correction of the deformities and prevention of further joint contractures. Physical and occupational therapy to stretch contracted joints, along with surgery and long-term bracing are the conventional modes of treatment (Eckel 1970; Lloyd-Roberts and Lettin 1970). These treatment modalities are facilitated by the physical improvement that takes place during the AMC patient's early developmental years. Although the physical presentation of the disorder may vary from limited mobility of the extremities to complete immobility, the patient's intelligence is generally not affected.

Published reports describe very few oral manifestations associated with this disorder. Main findings may include oral micrognathia, tongue protrusion, and limited mandibular movement (Pearson and Fowler 1963; Garfunkel and Steiner 1971; Sakamoto et al. 1985). Cohen and Isaacs (1976) noted TMJ involvement with associated trismus in 14 of 37 AMC patients.

Case Report

A 16-month-old white female with a diagnosis of AMC reported to the Marquette University School of Dentistry Special Patient Clinic for the placement of a mouthstick prosthesis. The patient's mother and an occupational therapist had noticed that the child had been using her nose and lips to turn pages in a book and move play objects. A mouth prosthesis had been suggested by the occupational therapist as an aid to helping the child become more independent in daily activities.

Medical Information

Following a pregnancy complicated by bleeding in the second trimester, the patient was born full term. Because she was a primary breech, delivery was done by Caesarian section. Her Apgar scores were 1-3-1-5 at the 1-, 2-, 5-, and 15-min intervals reflecting the severe respiratory distress she experienced. Her birth weight was 5 pounds, 12 ounces. She was at once noted to have arthrogryposis at all points as well as other congenital anomalies. The arthrogryposis was thought to result from neurogenic or myopathic origin since chromo-

some studies revealed a normal pattern.

Musculoskeletally there were marked contractures of all major joints with asymmetrical upper and lower extremities and marked muscle atrophy. The patient had limited control of her upper and lower extremities. Due to muscle agenesis, the patient could hold her head up briefly, but had trouble holding it in the midline.

The patient was considered to have a normal level of intelligence and verbal communicative ability consistent with her age.

Dental Examination

The patient was accompanied by her mother and occupational therapist and was cooperative during the examination. In order to provide support for her head/neck muscles and joints, the patient recently had been fitted with a body brace. This allowed her to sit in an upright position, providing guidance to support head movements. She was noted to have left-sided facial asymmetry with a normal mandibular range of motion.

Intraorally soft tissues were within normal limits. The primary dentition was present with the exception of the second primary molars. The maxillary canines were not fully erupted. No carious lesions were noted and oral hygiene was good. None of the oral manifestations reported in the literature to be associated with AMC were noted.

Materials and Methods

Because of the high level of patient cooperation and the strong support for learning from the patient's family and occupational therapist, a decision was made to fabricate the mouthstick prosthesis.

The mouthstick prosthesis was given to the patient 3 months after the initial dental appointment. During this period of time, the maxillary canines erupted, providing more occlusal stability and decreasing the need for early adjustments following mouthstick placement. The occupational therapist used this time to teach the child how to use the body brace to improve active control of head and neck muscles. This control enabled the child to direct the stick and tip portion of the prosthesis more effectively (Fig 1). The child also was taught how to hold objects between her teeth for long periods of time. This exercise was designed to simulate the mouthpiece utilization.

The prosthesis fabrication began with compound impressions of the existing maxillary and mandibular dentition. Compound rather than alginate was used as impression material due to its ease of handling and the decreased risk of the patient's aspirating the material. The process of fabrication was developed with consideration given to mouthstick prosthesis standards developed by Blaine and Nelson (1973).

FIG 1. Patient in body cast directing mouthstick prosthesis toward mother.

The maxillary and mandibular stone models were trimmed in order to improve adaptation of the wax which simulates the mouthpiece portion of the mouthstick prosthesis. The casts were mounted on a hinged articulator and the vertical dimension was increased 2 mm to allow for wax thickness. Next, the layer of baseplate wax was placed in a horseshoe fashion over the occlusal one-third of the mandibular teeth. The maxillary cast was then brought into the layer of wax so as to create an occlusal record of the maxillary teeth. The mouthstick prosthesis would only contact the occlusal portion of the teeth, thereby eliminating soft tissue trauma while the prosthesis was in function. A 1/4-inch screw was embedded into the wax anterior to the maxillary incisors. The threaded portion of the screw projected out from the wax. Once the wax mouthpiece portion of the prosthesis was fabricated, it was heat processed with high-impact resin material.

The stick portion of the prosthesis was fabricated from a lightweight, hollow, aluminum arrow shaft. Into the end of the aluminum stick portion, a plastic anchor was tapped into place. The plastic anchor accepted the threaded portion of the screw which was embedded in the mouthpiece. In this way the mouthpiece and mouthstick could be fastened together (Fig 2, next page). The length of the stick portion of the prosthesis was determined by the task requirements of the patient following discussion with her parent and occupational therapist. The aluminum stick was tilted 20° down to allow for better visualization by the patient. In order to facilitate the child's learning, a mouthstick prosthesis was also fabricated for the occupational therapist. This way the patient could learn how to use the mouthstick through practical demonstration by her therapist.

The mouthstick prosthesis was given to the patient and minimal adjustment was needed. The patient utilized the mouthstick prosthesis immediately and devel-

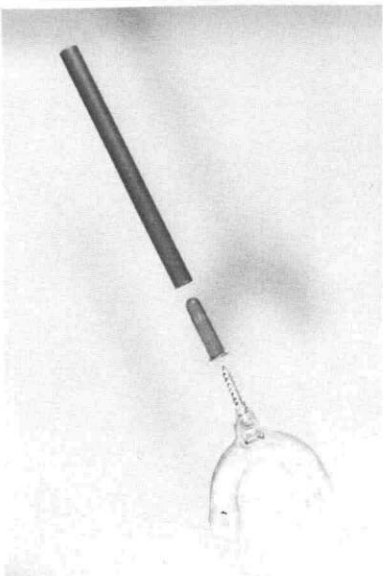

FIG 2. Mouthstick component parts as they fit together: arrow shaft, plastic anchor, screw, acrylic mouthpiece.

oped excellent skill through the demonstration of its use by the occupational therapist (Fig 3). The patient returned for follow-up appointments at 1 week, 2 and 6 months. It is anticipated that the patient will receive a new mouthstick prosthesis once the primary second maxillary and mandibular molars have erupted fully.

Discussion

AMC is a rare disorder that causes great difficulty for the afflicted individual due to its effects on joint mobility. The young child with this disorder is further limited in his/her developmental stages by being unable to manipulate objects while learning about his/her surrounding environment.

The mouthstick prosthesis is an excellent adjunct in helping persons with limited joint mobility to function in their environment. It can be fabricated by any pediatric dentist at low expense to the patient with readily available materials. Further, by also fabricating a similar prosthesis for the child's occupational therapist, learning how to use the mouthstick prosthesis is enhanced.

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FIG 3. Occupational therapist demonstrating use of mouthstick prosthesis to patient.

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