Congenital Central Hypoventilation Syndrome (CCHS) is a rare syndrome characterized by chronic respiratory failure in the absence of primary pulmonary, cardiac, neuromuscular, or chest wall disease. From birth, there are prolonged periods of apnea and hypoventilation, particularly during sleep. This usually results from a decrease in tidal volume (intake) as opposed to decreased frequency of respiration. After an initial period of mechanical respirator dependency, these patients usually develop the ability to sustain normal gas exchange while awake. However, ventilator dependency during periods of sleep persists as a result of the patient's inability to detect and react to potentially lethal levels of hypoxia and hypercapnia. There has been only 1 reported case where a child with this syndrome was able to attain normal gas exchange values during both waking hours and sleep and, thus, the need for mechanical ventilation was eliminated.

Children with CCHS are typically tracheostomized within the first few weeks of life. As they grow, they are monitored carefully to ensure that their tracheostomy tubes are of adequate size to prevent excessive air leakage during artificial ventilation. They typically demonstrate good psychomotor skills, but may have mild to moderate learning disabilities or cognitive skills. They often have problems with speech development because of inadequate air flow over the vocal folds when the tracheostomy tube is not covered or closed. This problem has been addressed in the past by using a tracheostomy tube of the same diameter as the one used for therapy, which has been shortened and plugged with silicone to prevent the escape of air through the stoma during speech. In addition, commercially available flanged silicone tracheostomal plugs can be fitted to the patient for this purpose.

As discussed by Ma and Ross in 1989 and Jacob and Bowman, various techniques have been used in the past to reproduce anatomic contours both of the peristomal and stomal areas. These techniques range from the modification of standardized laryngectomy tubes to using pediatric endotracheal tubes with an inflatable cuff to impress 2 to 3 inches beyond the stomal opening.

In this clinical report, a simple procedure for fabrication of a custom tracheostomal obturator is presented.

**CLINICAL REPORT**

A 10-year-old white girl with CCHS that was diagnosed within the first 3 weeks of life was evaluated for treatment. The patient was tracheostomized at 6 weeks of age and was ventilator-dependent during sleep. She appeared normal psychosocially, although a bit...
reserved. She was referred to the maxillofacial prosthetics service for fabrication of a custom tracheostomal obturator. Attempts at using pre-formed flanged tracheostomal plugs were rejected by the patient because the flanges were uncomfortable to her when the pros thesis was in place or removed. To effect speech, she would cover the end of her tracheostomy tube during which time air could clearly be heard to escape.

PROCEDURE

During initial examination, the size of the stomal opening was determined and recorded. The desired depth of the stomal obturator was determined by discussion with the patient’s otolaryngologist who was present. This depth was later tested to determine whether it was comfortable to the patient and did not interfere with airflow. A custom impression tray was constructed from base plate wax (TruWax, Dentsply/York Division, York, Pa.) and processed into clear acrylic resin (Hygenic clear laboratory resin, Hygenic Corp, Akron, Ohio) to the following specifications: (a) external flange and handle: $38 \times 25$ mm oval with solid external handle; (b) Perforated stem $31 \times 7 \times 2$ mm. Length includes terminal bulb. Similar angulation as tracheostomy tube; (c) terminal bulb diameter of the external stomal opening.

The patient returned to the Department of Otolaryngology/Head and Neck Surgery and, with the otolaryngologist present, the impression tray was tried in place to determine resistance to removal and relative comfort to the patient. The patient was able to breathe freely through the mouth and nose with comfort. The internal surface of the external flange, the stem, and the bulb were painted with Express polyvinyl siloxane (PVS) adhesive (3M, St Paul, Minn.) and then allowed to dry.

Equal amounts of Express PVS putty catalyst and base paste were mixed together and adapted to the terminal bulb and stem of the custom impression tray, ensuring that the perforations within the stem were well engaged. The impression tray was firmly seated in the stoma, then gently removed and worked back into place a second time to ensure passive fit. The patient then was instructed to perform functional movements of the head and neck (Fig. 2). Once set, the impression was removed and light-body PVS was syringed to the undersurface of the external flange and reseated. The patient repeated functional movements of the head and neck to capture an accurate impression of the external tissue during movements. The resultant
prosthesis is illustrated in Figure 1 adjacent to a pre-formed tracheostomy tube. The curvature of the prosthesis is similar to that of the pre-formed tube.

A tentative color match was performed using medical adhesive A (Dow Corning, Midland, Mich.) and silicone paste color concentrates dispersed in liquid silicone (Factor 11, Lakeside, Ariz.). This was used later as a guide for coloration of the silicone prosthesis. The impression was embedded into a vacuum mixed dental stone (Lab Stone-Buff, Modern Materials, Port Washington, N.Y.) in a disposable paper cup. The resultant mold was then scored and split and the impression was recovered and the mold reassembled.

Two flanges 4 mm in length each were added to the terminal bulb of the recovered impression. At this time, a second mold was poured. This was also scored and the impression recovered. Tops of both molds were smoothed with sandpaper and baseplate wax was added to their tops to form external flanges $38 \times 25 \times 2$ mm. The top of each mold was notched, and a cap was poured to fit each. The wax was boiled out and the molds allowed to cool. COE-Sep tinfoil substitute (GC America, Chicago, Ill.) was painted to all surfaces of each mold that would come into contact with the prosthesis (Fig. 3) so that it could be easily removed from the mold when completed.

Medical adhesive A was mixed with pigments to attain a color match with the patient’s skin tone in proportions that were determined previously. This mix was applied to the molds that were assembled and allowed to cure. The resultant prostheses were removed from their respective molds and trimmed.

A beaded necklace was assembled with parts from a local crafts store. The rear of the central bead was flattened, and the matrix portion of a clothing snap assembly was secured into place using autopolymerizing clear orthodontic resin (Fig. 4). The corresponding matrix portion of the snap assembly was secured with ligature wire to a small square of metal meshwork. A recess was cut into the center of the external surface of the prosthesis with a scalpel blade approximating the size of the metal mesh containing the matrix portion of the snap.

A second batch of Medical adhesive A and pigment was mixed in the same relative amounts as the first batch. This was used as a luting and a camouflaging agent, securing the metal mesh to the center of the prosthesis and disguising it to the eye. At the delivery appointment, the necklace was fitted to the patient with the prosthesis in place (Fig. 5). It was determined that the retentive flanges were not necessary for this patient. Not only were the flanges a source of irritation to the patient, but also because this prosthesis was resistant to removal with only the bulb in place. The necklace provided additional retention. On coughing, the prosthesis was secure, and there was virtually no air escape during speech.

**SUMMARY**

This clinical report describes the treatment of a patient with Congenital Central Hypoventilation Syndrome using a simple impression procedure and prosthesis fabrication. Treatment required only the basic information provided by the otolaryngologist, measurements were taken at the initial appointment, and basic skills were used in fabrication of intraoral and extraoral prosthetic devices. It is recommended that the clinical phase of this procedure be performed in the presence of an otolaryngologist. Patients who live with this syndrome have undergone many trials physically, emotionally, psychologically, and socially. The presence of this type of prosthesis has many advantages such as allowance of speech without obvious air leakage, and being relatively inconspicuous. Compared with the
alternative, this type of prosthesis allows these patients to feel less conspicuous.

REFERENCES


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Noteworthy Abstracts of the Current Literature

Effects of cyclic loading on the strength of all-ceramic materials

Purpose. This article evaluated the effects of fatigue on materials used for all-ceramic crowns. Biaxial flexural strength of the all-ceramic materials were measured with prepacked and laminated specimens after cyclic loading.

Material and methods. Specimens were prepared from 2 types of all-ceramic systems, a glass-infiltrated alumina core system (In-Ceram) and a leucite-reinforced feldspathic porcelain system (IPS-Empress). Monolayer and laminated disks (11.75 mm diameter, 1.2 mm thickness) were prepared. Biaxial flexural strength of the specimens (n = 10 for each group) that were polished and created with a precrack were measured. Their strength after cyclic loading was also measured. A cyclic load that was 60% of the mean break load of the specimens was applied (before cyclic loading) to specimens for $10^5$ cycles in 37°C water. Results were statistically evaluated using ANOVA and Student’s t test.

Results. Although 20% to 30% of the polished specimens samples fractured during cyclic loading, biaxial flexural strength of specimens that survived was approximately the same as that of specimens not subjected to cyclic loading. Strength of the alumina system decrease with the introduction of precracks. Nearly all specimens fractured during cyclic loading. Strength of the leucite system did not decrease with the presence of precracks and no fractures were observed during cyclic loading.

Conclusion. The results suggest that although the alumina system has high flexural strength, it was more sensitive to flaws and to fatigue fracture. The effect of fatigue on the leucite system appeared to be low. 29 References. — R. P. Renner