Pierre Robin sequence (PRS) was first described in 1923 by the French dentist Pierre Robin and occurs in one of 8500 to one of 14,000 births. PRS is characterized by micrognathia, glossoptosis, and airway obstruction. It is usually associated with a palatal cleft, which is not required for diagnosis. PRS is not defined as a syndrome, but a sequence in which each anomaly results from a previous one in a sequential chain. In this regard, micrognathia leads to glossoptosis, which subsequently results in airway obstruction and feeding problems.

Respiratory insufficiency is the most severe complication of PRS. The degree of airway obstruction in PRS is variable. Some infants can obtain their airway when they are awake but suffer from obstruction when they are asleep. This is enhanced in the supine position. Severe cases need surgical interventions for airway management. Tracheostomy should be avoided because a relevant proportion of cases cannot be successfully weaned. If conservative measures are not sufficient, early distraction should be considered. Mandibular distraction osteogenesis was first described by McCarthy in 1992. The advancement of the mandible by 15 mm could be achieved within 2 weeks. Thereafter, the small patient could be extubated successfully, and there was no further episode of major respiratory insufficiency. We demonstrate that mandibular early distraction with a patient-specific distractor is a successful method to treat severe respiratory insufficiency in PRS, and it can prevent the necessity for tracheostomy with the resulting disadvantages. We provide details concerning our therapeutic algorithm, metric analyses, and a discussion of relevant literature.

CASE PRESENTATION

We report the case of a female neonate with PRS, a prenatally confirmed chromosome 4q deletion syndrome, in combination with cardiac insufficiency. After delivery in an external hospital (birth weight, 2940g; height,
the newborn showed repeated episodes of respiratory insufficiency. Placing the newborn in prone position was not sufficient. A Tuebingen airway plate was inserted but not tolerated; so the newborn had to be switched to Continuous Positive Airway Pressure (CPAP) ventilation.

The 18-day-old patient was transferred to the pediatric cardiac surgery unit of our university hospital for treatment of an unbalanced atrioventricular septum defect and hemodynamically relevant patent ductus arteriosus leading to increasing cardiac insufficiency. The next day, extraluminal central pulmonary artery banding and ductal ligation were performed. Endotracheal intubation for this procedure was extremely difficult. After induction of anesthesia, intubation was attempted in the first instance using conventional direct laryngoscopy, showing a Cormack Lehane IV-grading. Therefore, in the second effort, intubation was performed successfully using a video-laryngoscope-assisted flexible bronchoscopic technique (hybrid technique). On the following day, the patient was extubated and transferred to CPAP ventilation. Due to a pronounced micrognathia with glossoptosis and cleft palate, an anatomically caused obstruction of the oropharyngeal airway had led to asphyxia and bradycardia with consecutive cardiac and circulatory arrest in the 21-day-old patient, which made cardiopulmonary resuscitation necessary. After 2 minutes of chest compressions, the patient could be intubated again using the hybrid technique, and we performed a glossopexia as an emergency measure.

The patient could be ventilated stably with mild ventilation parameters and was sedated with fentanyl and clonidine. Due to the extreme glossoptosis, no extubating attempt was performed despite the glossopexia, and the interdisciplinary decision was made for early mandibular distraction osteogenesis. Four days later, a computed tomography (CT) scan was performed for virtual surgery planning. Endotracheal intubation was prolonged until surgery.

Virtual surgery planning of the mandibular distraction [Computer Aided Design/Computer Aided Manufacturing (CAD/CAM)] of 15 mm (Figs. 1 and 2) was performed together with KLS martin (Tuttlingen, Germany). At the age of 35 days, two internal patient-specific distractors were placed at the mandibular angle using a submandibular incision. From the first postoperative day, the distractor was activated twice daily for 15 days to achieve the planned

**Fig. 1.** Preoperative CAD/CAM planning of the distractor positioning. A, The 3D renderings show the preoperatively planned insertion position of two internal patient-specific distractors at the right and left mandibular angles. A commercially available virtual surgery CAD/CAM planning together with our industry partner KLS martin (Tuttlingen, Germany) was performed. The position of the mandibular nerve and the tooth follicles is marked. B, Renderings show the osteotomy lines of the left and right sides and screw positions in relation to the mandibular nerve and tooth follicles.

**Fig. 2.** Preoperative CAD/CAM planning of the distractor activation. Renderings show the final mandibular position after full activation of the distractor. Mandibular bone is indicated in red. The bone gap shows the intended movement of 15 mm.
distraction distance of 15 mm. A CT scan was performed to assess the result. Twenty days after distractor placement, the patient was extubated successfully.

The distractors were removed 13 weeks after insertion. Early mandibular distraction showed a satisfactory functional and aesthetic result. Volumetric assessment revealed an improved mandibular position and upper airway width (Figs. 3 and 4).

**DISCUSSION**

Prone or lateral position relieves airway obstruction of PRS affected infants in approximately 70% of cases. If this is not sufficient to resolve the airway insufficiency, an attempt can be made to bypass the airway obstruction with a nasopharyngeal tube. Surgical airway management procedures include glossopexia, distraction osteogenesis, and tracheostomy. The principle of glossopexia is to pull the tongue forward and suture it to the lower lip. Success rates of over 80% are described in the literature for this technique. In our case, nasopharyngeal tube and glossopexia did not improve respiratory insufficiency.

The literature considers neonatal mandibular distraction osteogenesis as a safe procedure. There are only a few prospective studies on the surgical treatment of PRS. One prospective study with 28 cases showed the necessity for mandibular distraction in 10 cases. There was no difference in time to discharge, need for gastrostomy, and weight gain. Tracheostomy could be avoided in all cases. A recent retrospective analysis of early mandibular distraction in 69 PRS infants reported high success rates, even in patients below 3 kg body weight. Distraction was performed at a mean age of 35 days, and tracheostomy was necessary in two cases. Analysis of 117 infants with PRS and early mandibular distraction at an average age of 71 days revealed an enlarged upper airway cross section, which was also shown in our case (Fig. 4).
analysis in 171 PRS cases revealed an association of necessity of tracheostomy with neurologic impairment.10

Alternatively, a stock distractor could be used instead of a custom fabricated one. This comes with the advantage of a faster time to surgery and lower costs. However, in the presented case, surgery had to be postponed anyway because of the need for cardiopulmonary stabilization after resuscitation. In contrast, the advantages of virtual surgery planning are reduced risk of injury of adjacent structures and reduced operating time.

CONCLUSIONS

Virtual planning for early mandibular distraction osteogenesis and the use of a patient-specific distractor can help obtain an optimal distraction vector, reduce operation time, and protect structures like tooth follicles and the inferior alveolar nerve. Treatment decisions should be made by an interdisciplinary team involving at least anesthesiologists, pediatricians, and maxillofacial surgeons.

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