Outcomes analysis of neonates treated for Pierre Robin Sequence: An 18-year experience.

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Background: Pierre Robin Sequence (PRS) is caused by micrognathia, which leads to glossoptosis and airway obstruction. Multiple treatment modalities are described including conservative treatment, bypass of the obstruction, airway repositioning and correction of the anatomic deficiency. This study presents an outcomes analysis of the largest series of neonates with PRS.

Methods: An IRB approved, 18-year retrospective review of all neonates treated with PRS was performed. Examined variables included patient demographics, syndromic and neurologic status, feeding outcomes, and polysomnography and microlaryngoscopy data.

Results: 140 neonates were identified and divided into three cohorts based on initial treatment: conservative management (n=22), external mandibular distraction (MD, n=68) and tracheostomy (n=50). Neonates initially receiving tracheostomy were more likely to be syndromic (OR=2.62, p=0.01), have neurologic impairment (OR=2.85, p=0.02), GE reflux (OR=2.1, p=0.05) and require intervention within 5 days of birth (OR=60.65, p=0.005) compared with those receiving MD. Polysomnograms obtained pre- and post-intervention had similar significantly improved profiles for patients receiving both MD (obstructive index (OI) decrease 43.8 (p=0.03)) and tracheostomy (OI decrease 44.7 (p=0.008)). However those receiving MD had significantly greater avoidance of gastrostomy (19.1 vs 83.7%, p<0.0001) and higher success in exclusive oral diet (85.7% vs 38.9%, p<0.0001) compared to the tracheostomy patients. As shown in Table 1, four factors were associated with failure of MD: low birth weight, syndromic status, neurologic impairment and poor post-intervention polysomnogram. We demonstrate that combined these factors may be used to predict those at risk for failure of MD with high sensitivity and specificity (Table 2). Lastly, the presence of multilevel obstruction identified on microlaryngoscopy was not associated with need for tracheostomy (38.6% vs 26.9% for MD group, OR 1.77, p=0.22); and when present in patients receiving MD multilevel obstruction was not associated with higher failure rates (p=0.71).

Conclusions: Mandibular distraction is an efficacious treatment modality for neonates with PRS, and should be considered the first line intervention to avoid tracheostomy. Patient variables including birth weight, syndromic status, neurologic impairment and multi-level airway obstruction do not preclude the utilization of MD; however, predicting the future necessity of tracheostomy and the ability to decannulate following MD of these patients is less reliable.

Table 1. Factors predicting failure of mandibular distraction for treatment of micrognathia/tongue-based airway obstruction. (Failure defined by need for subsequent tracheostomy.)

	MD success (%) 62 (92.2)	**MD failure (%) 6 (8.8)**	**Odds Ratio**
Birth weight <2.5kg	14/58* (24.6)	4/5 (80.0)	12.57 (p=0.03)
Syndromic status	23/62 (37.1)	5/6 (83.3)	8.48 (p=0.05)
Neurologic impairment	12/53 (22.6)	4/6 (66.7)	8.33 (p=0.02)
Post-MD sleep study with Respiratory Disturbance Index <20	33/36 (91.7)	2/4 (50.0)	11.00 (p=0.04)

*Not all data points were available for each patient but all available points are presented.

Table 2. Mandibular distraction failure prediction testing, based upon the four criteria in Table 1.

	3 criteria available * 2 criteria present	3 criteria available* 3 criteria present	4 criteria available All 4 criteria present
MD Success (n=62)	12/51	3/51	0/31
MD failure (n=6)	5/5	3/5	2/2
Test sensitivity	100%	60%	100%
Test specificity	76%	94%	100%
Positive predictive value	29%	50%	100%
Negative predictive value	100%	96%	0%

* The three criteria used here are low birth weight (<2.5kg), syndromic status and neurologic impairment. Only patients for whom the three criteria (or four criteria for the right column) were available were included.