INTRODUCTION

The definition of Pierre Robin sequence (PRS) used in this paper is the triad of micrognathia, glossoptosis, and cleft palate. The sequence is recognized to occur in approximately 1 in 8000 live births, up to 80% of which have an additional syndrome.1,2

Respiratory obstruction and feeding difficulty can present with a wide range of severity in patients with PRS. In patients with severe airway compromise, tracheostomy was previously considered conventional management.3 Since the early 2000’s, however, mandibular distraction osteogenesis (MDO), an accepted method among otolaryngologists and craniofacial surgeons alike for alleviating airway obstruction, avoiding tracheostomy, and restoring feeding ability in infants with PRS,4-11

Many institutions consider computed tomography (CT) a standard step for pre-operative planning of MDO. Indications cited for pre-operative CT include planning for osteotomies and vectors of distraction, locating the inferior alveolar nerve and inferior tooth buds, and assessing the temporomandibular joint (TMJ).9 The authors of this paper do not routinely obtain pre-operative CT for infants with isolated PRS or Stickler syndrome prior to MDO, given potential downstream effects of radiation exposure to the head and neck area in infants.12-15 It is important to note that we only use externally applied, multi-directional distraction devices in this patient population.

METHODS AND MATERIALS

Infants who underwent MDO between 1998 and 2014 were identified from databases at two medical centers using procedure codes. Inclusion criteria were isolated PRS (iPRS), age less than nine months at time of MDO, and use of external distraction devices. Patients with Stickler syndrome were included as well, as these patients resemble iPRS infants and are often not diagnosed with Stickler syndrome after the neonatal period if not years later.

RESULTS

Fifty-two patients with iPRS / Stickler syndrome underwent MDO; 42 did not receive pre-operative imaging. Mean age at distraction was 35 days (6 days -7 months). Follow up ranged from 5 days to 8 years. All 42 patients without imaging successfully avoided tracheostomy or achieved decannulation. Two patients (4.8%) required post-operative gastrostomy tube placement. Two patients (4.8%) had minor intraoperative complications that might have been anticipated with preoperative CT. Two patients (4.8%) demonstrated malocclusion at the end of the distraction phase; asymmetry occurred in one patient (2.4%). There were no differences in outcomes between CT and non-CT groups in five out of seven outcome measures (Table 1).

CONCLUSIONS

This series suggests that preoperative CT imaging may not be necessary to achieve consistently successful outcomes following mandibular distraction osteogenesis using multidirectional external distractors in infants with iPRS or Stickler syndrome. This has implications for cost containment and improved patient safety through reduction of medical imaging radiation exposure.

REFERENCES


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