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Long-term orthognathic surgical outcomes in Treacher Collins patients

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Summary *Introduction:* Treacher Collins syndrome is a rare disorder characterized by several orofacial findings including malar deficiency and hypoplastic mandibles. These patients often require a combined orthodontic—orthognathic approach to correct their malocclusion. This is most often characterized by a short posterior vertical height and an anterior open bite. Orthognathic correction often requires Le Fort I and bilateral sagittal split osteotomies. No long-term stability results have been reported after bimaxillary surgery in Treacher Collins patients.
Methods: A retrospective review of all Treacher Collins patients evaluated for orthognathic surgery by a single surgeon from 1993 to 2007 was performed. Patients were divided into groups who required surgery and those who did not. Part I analyzed the cephalometric differences between the surgical (S) and nonsurgical (NS) groups. Part II of the study assessed the preorthodontic treatment (T_1), preoperative (T_2), immediate postoperative (T_3), and 1-year postoperative (T_4) cephalometric measurement variables to determine the net surgical movement ($T_3 - T_2$) and relapse ($T_4 - T_3$).
Results: Twenty-two patients met the inclusion criteria, of which 11 had occlusal relationships requiring orthognathic surgery. Nine out of 11 chose to have surgery.

At baseline, surgical patients exhibited a statistically significant retruded maxilla as measured by SNA and midface length compared to the NS group. In addition, the S group also had an increased gonial angle. There were significant movements in all maxillary and mandibular measurements. There was a significant relapse in the palatal plane angle when the maxilla was anteriorly impacted, with a 2.8-mm average relapse of the advancement. Relapse of the counterrotation movement of the mandible was identified, but this was not significant. Relapse did not affect the final occlusal result, which may have been compensated with postsurgical orthodontic treatment.
Conclusion: Bimaxillary orthognathic surgery in the Treacher Collins patients may be performed safely with long-term dental and skeletal stability.

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Introduction

Treacher Collins syndrome is a rare autosomal dominant syndrome affecting the first and second branchial arches, first described by Edward Treacher Collins in 1900.[1,2] The reported incidence is one in 25,000—50,000 live births.[3] The clinical features of Treacher Collins syndrome consist of a characteristic appearance including mandibular micrognathia, malar deficiency, down-slanting eyes, and euryblepharon with or without coloboma.[4] At the severe end of presentation, there is a complex orbitomalar—zygomatic cleft and class II malocclusion typified by an anterior open bite. A prominent nasal dorsum with a clockwise-rotated mandible (as viewed on a standard cephalogram from the right side) gives a convex profile to the face.[2,5,6] In 28% of patients, maxillary hypoplasia may be accompanied by a cleft palate.[7] This is thought to be secondary to a restricted tongue position within a diminutive oropharynx, causing glossoptosis over a retrognathic mandible. Nasopharyngeal volume is also reduced and may be accompanied by choanal atresia, with resultant airway issues that present early in the newborn.

Patients presenting with Treacher Collins syndrome at the Hospital for Sick Children Centre for Craniofacial Care and Research are evaluated by a multidisciplinary team assessment. These include dental, speech, orthodontic, and plastic surgical consultations. Procedures such as bimaxillary advancement, rhinoplasty, genioplasty, and soft tissue augmentation can deliver a significantly improved facial aesthetics and quality of life when delivered with a multidisciplinary approach to care.[8—11] Bimaxillary surgery can restore facial balance and level the occlusal plane. These surgical movements can be technically challenging to achieve in patients with Treacher Collins syndrome due to poor bone stock and soft tissue deficiency. Previously, we reported that the mandible and midface in Treacher Collins patients are expectedly short in the anteroposterior plane compared to controls.[11] The mandibular plane angle is obtuse, affecting female patients in particular, in whom there is also a posteriorly placed chin point. The affected individuals have a high antegonial notch, which increases the complexity of fixation. Although anterior lower facial height proportion in the control and syndromic patients are similar, the posterior facial height in Treacher Collins syndrome patients is reduced.[11] Due to the shortened posterior facial height in Treacher Collins patients, the orthognathic movements routinely include an anterior maxillary impaction with posterior maxillary extrusion. These movements result in correction of the palatal plane angle and opening of the posterior nasopharyngeal airway. Leveling of the maxilla results in an even greater rotation of the already obtuse mandibular plane during the sagittal split. Even with these complexities, bimaxillary orthognathic surgery in Treacher Collins patients improves facial balance and projection, lessening the stigmata of the disease.

To date, there have been no long-term reports of bimaxillary orthognathic surgery in Treacher Collins patients. There have only been case reports that suggest mandibular relapse after 1—2 years following mandibular surgery in patients with Treacher Collins Syndrome.[12,13]

The purpose of this study is to examine the long-term stability of bimaxillary orthognathic surgery in a Treacher Collins population. It is important to counsel patients and their families effectively on the expected outcomes.

Methods

A retrospective chart review was performed on patients diagnosed with Treacher Collins syndrome at the Hospital for Sick Children from 1993 to 2007. Patients were included only if they had Treacher Collins syndrome confirmed as a diagnosis by a clinical geneticist, a combined orthodontic—orthognathic treatment, pretreatment with lateral cephalometric radiographs, pre-surgery, immediately postop, and at least 1 year post-op. Orthognathic surgery was performed by the senior author (JHP). Patients were excluded from the study if they had any other genetic diagnosis in addition to Treacher Collins syndrome, a history of distraction osteogenesis, or were undergoing active orthodontic treatment.

The study was divided into two parts. Part I of the study analyzed cephalometric differences between the surgical (S) and nonsurgical (NS) Treacher Collins groups. Jaw position, midface and mandibular length, facial plane angles, facial height ratios, and antegonial notch height were recorded (Figure 1). Data were obtained from lateral cephalograms and analyzed using the registered software Dentofacial Planner 7.2 (Dentofacial Software Inc., Toronto, Ontario, Canada). All cephalometric data were collected and analyzed by a single trained user (MCC).

Part II of the study analyzed the surgical group using the same cephalometric parameters. For each parameter, there was a pretreatment (T1), presurgical (T2), immediate postoperative (T3), and at least 1-year-postoperative (T4) measurement (Figure 2). The surgical movement was calculated as T3 − T2. The relapse was calculated as the change from T3 to T4 (T3 − T4), and the total gain was calculated as the surgical movement minus relapse (T4 − T2). Student's *t*-test for paired parametric data was applied to obtain values for statistical differences across time for the datasets. Single measurements were recorded for each patient, and the standard deviation represents the variation within the patient population.

Results

Sixty-seven patients were identified over the study period with a diagnosis of Treacher Collins syndrome. Of these, a total of 22 patients met the inclusion criteria with an appropriate follow-up. There were 13 male and nine female patients. Of the 22 patients, 11 were deemed to have occlusal relationships that required orthognathic surgery for correction. All patients had a class II malocclusion relationship. Eleven patients had functional occlusion or occlusions that could be treated with orthodontics alone. Of the 11 cases warranting surgery, nine patients underwent bimaxillary surgery, while two did not wish to pursue surgical treatment. Of the 11 patients who did not undergo orthognathic surgery, three patients had some other form of surgical intervention such as malar augmentation, fat injections, bilateral canthopexies, or genioplasty. All