The Apert and Crouzon syndromes: General and dental aspects

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This thesis is based on the following papers:

I. Hallberg U, Stavropoulos D, Mohlin B, and Hagberg C. (2011): ***Living with Crouzon syndrome: Transition from childhood to adulthood.*** *Scand J Disabil Res* In press

II. Stavropoulos D, Hallberg U, Mohlin B, and Hagberg C. (2011): ***Living with Crouzon syndrome: How do young adults with Crouzon syndrome handle their life situation?*** *Int J Paed Dent* 21: 35-42

III. Stavropoulos D, Mohlin B, Kahnberg K-E, and Hagberg C. ***Comparing patients with Apert and Crouzon syndromes: Clinical features and cranio-maxillofacial surgical reconstruction.*** Submitted

IV. Stavropoulos D, Bartzela T, Mohlin B, Kahnberg K-E, and Hagberg C. ***Dental agenesis patterns in Crouzon syndrome.*** Submitted

V. Stavropoulos D, Bartzela T, Bronkhorst E, Mohlin B, and Hagberg C. (2011): ***Dental agenesis patterns of permanent teeth in Apert syndrome.*** *Eur J Oral Sci* In press

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Abstract

Background: Craniofacial malformations, as seen in Apert and Crouzon syndromes, may have an immense impact not only on function and esthetics, but also on the psychosocial well-being of the person affected.

Aims: To provide insight on the social life aspects of persons with Crouzon syndrome in Sweden, during the transition from childhood to adulthood and as young adults. Furthermore, to study the main facial and intraoral characteristics of persons with Apert or Crouzon syndrome, the clinical manifestations that may be present in addition to the main syndromic features, and the cranio-maxillofacial surgical treatment protocols followed. Finally, to investigate dental agenesis and dental agenesis patterns of permanent teeth in persons with these syndromes.

Material and Methods: Firstly, interviews according to the qualitative method of Grounded Theory were carried out. Eight persons with Crouzon syndrome participated. Then, 23 patients with Apert syndrome and 28 patients with Crouzon syndrome were evaluated for general aspects, craniofacial aspects, dentoalveolar traits before and after the final orthognathic surgery, types and timing of cranio-maxillofacial surgical operations. Finally, dental agenesis and dental agenesis patterns were studied in 26 persons with Crouzon syndrome and in 23 individuals with Apert syndrome by evaluation of serial panoramic radiographs.

Results and Conclusions: The analysis of the interviews revealed that persons with Crouzon syndrome had to face different obstacles when developing their self-image during the transition from childhood to adulthood. Young adults with Crouzon syndrome tried to make the best of their situation. Already from childhood, they developed various strategies that helped them to cope with their lives. Mental disability, associated additional malformations, cleft palate, and extensive lateral palatal swellings were more common in children with Apert syndrome. In both syndromes, clinical findings included concave profile, negative overjet, posterior crossbites, anterior openbite, and dental midline deviation, which were significantly improved in almost all instances after the final combined orthodontic and orthognathic surgical treatment. The only exception was the posterior crossbites, which were persisting in about half of the cases. Cranial vault decompression and/or reshaping, midfacial and orbital advancement procedures, often in conjunction with a mandibular set-back, were the most frequent cranio-maxillofacial operations performed in both of the syndromes investigated. The prevalence of agenesis for at least one tooth was 42.3% for the patients with Crouzon syndrome. The dental agenesis patterns showed a remarkable variability. The prevalence of agenesis for at least a tooth was 34.8% for the patients with Apert syndrome. Symmetrical and repetitive dental agenesis patterns were identified.

Key words: Apert syndrome, Crouzon syndrome, social life, grounded theory, clinical features, cranio-maxillofacial surgery, dental agenesis, dental agenesis patterns

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