

Associated orthodontic, surgical and hematological management of Cooley's anemia

Report of a case

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Cooley's anaemia or β -thalassaemia or mediterranean anaemia is a blood disease characterised by malformation of the skull and long bones, which confers a typical appearance on the patient. The complete development of facial abnormalities can be prevented by an intensive blood transfusion programme or by bone-marrow-transplantation. At the present time these therapeutic strategies would be able to help these patients grow and develop, live a prolonged life and avoid bad surgical results. The aim of this study was to evaluate the feasibility of orthodontic and maxillofacial surgical corrective treatment associated with an appropriate transfusion therapy in a β -thalassemic patient. The patient enrolled in the study was affected by major β -thalassemia and diagnosis was performed as an infant. She was referred to our centre at puberty for dento-maxillofacial disorders. Clinical, haematological and radiographic evaluation permitted a complete diagnosis. She received a combined haematological, orthodontic and maxillofacial surgical treatment. Controls for evaluating the statement of results obtained were performed at different times after the end of the therapy and have shown that the therapeutic objectives had been achieved and maintained. At the present time, complete diagnostic and therapeutic haematological strategies cannot be carried out on a large scale, especially in coun-

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tries where health resources are limited. This objective reason, associated with possible low patient compliance, explains why we still encounter severe facial deformities resulting from erythroid hyperplasia. Our results suggest that this facial disfigurement requires surgical and orthodontic correction by consolidated surgical-orthodontic techniques performed according to the appropriate anaemia therapy. Although this is a preliminary study, initial encouraging results show no relapse three years after the end of the therapy.

Key words: β -Thalassemia, therapy - Craniofacial abnormalities, surgery - Orthodontics - Blood transfusion.

About 3% of the world's population, corresponding to 150 million people, carry β -thalassaemia genes.¹ These genes are particularly prevalent in inhabitants of Italy and Greece. The highest prevalence of the carrier state has been found in Sardinia, ranging from 11% to nearly 35%.

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