

The Esthetic management of pediatric patient with a hereditary disease (Schwachman–Diamond syndrome)

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Abstract

The Schwachman–Diamond syndrome is an autosomal recessive syndrome(1/20,000 births), consisting of pancreatic insufficiency, neutopenia, which may be intermittent, neutrophil chemotaxis defects, metaphyseal dysostosis, failure to thrive and short stature. Patients present in infancy with poor growth and grease, foul–smelling stools that are characteristic of malabsorption.

These children can be readily differentiated from those with cystic fibrosis by their normal sweat chloride levels, lack of the cystic fibrosis gene, and characteristic metaphyseal lesions. Pathologically, the pancreatic acini are replaced by fat with little fibrosis.

The neutropenia may be cyclic. Recurrent pyogenic infections otitis media, pneumonia, dermatitis(fig 1), sepsis are common and a frequent cause of death.

In dental examination, these patients had a poor oral hygiene and moderate generalized marginal gingivitis, also show delayed primary tooth exfoliation and oral development.

This report illustrates a case that pancreatic agenesis 6 yeas–old boy with various esthetic dental problems has been served the esthetic dental restoration of 6 years.

General & Dental examination

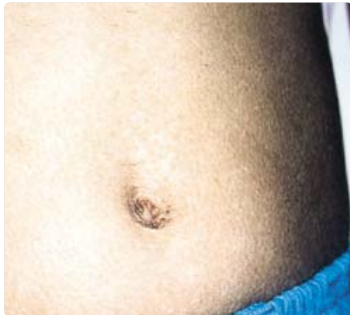
In first visit, that child, 6 years old male, had developmental delay with IQ assessed as below average, and he had generalized characters of the Schwachman– Diamond syndrome.(fig 1,2)

His weight was 11kg and height was 98cm.(fig 3) 6 years later, at 12 years old now, The weight increased about 6kg and height did about 12cm.(fig 4)

Dental examination included poor oral hygiene, severe dental caries on primary molars, moderate generalized marginal gingivitis, generalized calculus deposit, delayed primary tooth exfoliation and hypoplastic enamel with wide spread pitted defects.(fig 4,5,6)

These dental and esthetic problem have been observed repeatedly from first vist to now.

Schwachman–Diamond syndrome



[fig 1] Dermatitis



[fig 2] The Bended legs caused by the nutritional deficiency



[fig 3] The height of patient at first visit in 1999 was 98cm, and his weight was 11kg



[fig 4] The height of patient at last visit in 2004 was 110cm, and his weight was 17kg

Treatment

In primary dentition, all deciduous molar were done pulp treatment or extraction, then cementation of S-S crown, Crown-loop space maintainer. In maxillary incisors were restored by Resin and Glass Ionomer.(fig 7,8)

Primary dentition(In first visit)



[fig 5] The caries lesion on primary ant. teeth [fig 6,7] The severe caries lesion on primary molars

In mixed dentition, they were done that space supervision and various orthodontic treatment to introduce proper occlusal development. Of course, continuous preventive dental tx and periodic scaling were done, too.(fig 9,10)

Treatment and Oral hygiene management from primary dentition to mixed dentition



[fig 8] SS crown and Space maintainer on primary molars

[fig 9] P&F sealant on newly erupted permanent molars

[fig 10] Diastema between central incisors and the improvement of periodontal tissue throughout total periodontic tx.

Then now, In permanent dentition, permanent molars with carious lesion were treated by various esthetic restorations, such as composite resin, and operated periodontal treatment to remove generalized calculus and pigmentation. Finally, preventive care consisting of regular professional prophylaxis, topical fluoride application should be to control his high susceptibility of dental caries and enamel breakdown during the period of tx.(fig 11,12,13)

Discussion

The Schwachman–Diamond syndrome is very painful disease because of its' gastroenterologic problem. The patient has the severe physical and mental retardation. So, many esthetic dental problem develop continuously.

In this case, The child with various esthetic and dental problems who had the Schwachman–Diamond syndrome was treated by esthetic restorations, orthodontic treatment, and preventive care. In present, he is 12 yrs old and his quality of life and esthetic were improved.(fig 13,14)

In permanent dentition



[fig 11] The occlusal view of Mandibular dentition in last visit



[fig 12] The occlusal view of Maxillary dentition in last visit



[fig 13,14] The ant. view and a ficial photo in last visit



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