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PEDODONTIC MANAGEMENT OF EHLERS-DANLOS SYNDROME

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ABSTRACT

Ehelr Danlos Syndrome (EDS) is characterized by hyperextensibility of skin, hypermobility of joints n resulting in dislocations, and tissue fragility. It is a clinically and genetically heterogeneous connective tissue disorder affecting 1 in 5,000 individuals. Heterogeneity of it makes accurate diagnosis difficult. Preventive and supportive treatment and regular monitoring of patient's oral and physical status may be helpful in treating this kind of patient. Pedodontists have important role in initial diagnosis and preventive as well as corrective dental care of EDS patients.

INTRODUCTION

Ehlers-Danlos syndrome (EDS) is an inherited heterogeneous group of connective tissue disorders characterized by hyperextensibility, hypermobility of joint and delayed wound healing affecting skin, ligaments, joints, blood vessels and other organs. In 400 BC Hipprocrates described it.[1] So it is supposed to be one of the oldest known causes of bruising and bleeding. In 1901 Edvard Ehlers, a Danish dermatologist recognized the condition as a distinct entity.[2] In 1908, Henri-Alexandre Danlos, a French dermatologist, suggested that skin extensibility and fragility were the cardinal features of the syndrome.[3] Danlos expressed peculiar cigarette paper scars and pseudotumour formation of the skin and also laid down four diagnostic criteria, namely, hyperelasticity of skin, fragility of skin, hypermobility of joints and subcutaneous molluscus pseudotumour formation. In 1934, Pomeau-Delille and Soulied named the condition as Ehlers-Danlos syndrome by.[4] In 1936 Parkes Weber described 3 cardinal features: hyperextensibility of the skin, looseness of the joints, and friability of the skin and blood vessels.[5] In Indian literature very few number of cases has been reported till date.[6],[7],[8] Management of pediatric dental problems associated with Ehlers-Danlos is difficult and needs proper evaluation, care and expertise.

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Types and clinical stomatognathic manifestations of EDS

Types - The different types of EDS are classified according to the signs and symptoms. The types of EDS are (1) Hypermobility (previously EDS Type III) Joint hypermobility is the dominant clinical manifestation. Chronic joint and limb pain is a common complaint among individuals. (2) Classical (Previously EDS Types I & II) Marked skin hyperextensibility with widened atrophic scars and joint hypermobility are found. (3) Vascular (Previously EDS Type IV) most serious form of EDS due to the possibility of arterial or organ ruptures. The skin is usually thin and translucent with veins being seen through the skin surface. This is most apparent over the chest and abdomen. There are certain facial manifestations like large eyes, thin nose, lobeless ears, short stature and thin scalp hairs present in some affected individuals. (4) Kyphoscoliosis (Previously EDS Type VI) General joint laxity and severe muscle hypotonia at birth are seen. The muscular hypotonia may be pronounced and leads to delayed gross motor development. Individuals with the kyphoscoliosis type present with progressive scoliosis at birth. (5) Arthrochalasia (Previously EDS Type VII A&B). Congenital hip dislocation has been present in all biochemically proven individuals. Severe generalized joint hypermobility with recurrent subluxations are seen. (6) Dermatosparaxis (Previously EDS Type VIIC). Have severe skin fragility and substantial bruising.

Clinical stomatognathic manifestations of EDS

Area		Clinical manifestations	Clinical Significance
Intraoral			
	Teeth	1. Enamel Hypoplasia.[9]	1. Enamel hypoplasia a. Need urgent evaluation. b. Preventive management. c. Esthetic management.
		2. Supernumerary teeth.[10]	2. Super numeracy a. Wait & observe. b. Rarely surgical intervention.
	Periodontal structure	1. Periodontitis[11]	1. Periodontitis needs proper evaluation & necessary periodontal management.
		2. Hemorrhage	2. Hemorrhage needs immediate arrest of bleeding.
		3. The fragility of the gingiva	3. Gentle tissue handling
		4. Premature loss of deciduous and permanent teeth.[12],[13]	4. Premature loss of teeth – a. Needs parental attention, b. Professional consultation.
		5. Bleeding on brushing	5. Gentle brushing with soft bristled tooth brush
	Oral mucosa	1. Easy mucosal tears during instrumentation. 2. Failure to retains Sutures.[16] 3. Reduced healing capacity	1. Gentle tissue handling 2. Pressure pack application 3. Needs proper post operative care
	Palate	Vaulted palate.[12],[14]	Professional consultation for Emergence of any mal occlusion.
	Tongue	Gorlin's sign	Patient motivation
Extra Oral	Skin	1. Scarring	Avoid further traumatic exposure Avoid enthusiastic
		2. Hyperelasticity	Demonsration of Hyperelasticity to the peer group.
	TMJ	1. Pain 2. Repeated luxations	1. Pain management. a. Precautionary measure to avoid large mouth opening. b. Avoid too much yawning.
	Ear	Prominent Pinna	No treatment needed
	Eye	Blue sclera	Professional consultation
	Forehead	Scar mark	Avoid further traumatic exposure
	Chin	Scar mark	Avoid further traumatic exposure

A 11-year-old male patient was referred to our Department for diagnosis and treatment. According to his patient history, he was born at week 38 of the pregnancy. His birth weight was 2750 g and length was 53 cm. His mother, a housewife, did not notice his muscle weakness at birth. His prelinguistic speech development was started at 6 Months of age, and his cognitive and intellectual development was normal. After3 years, further physical symptoms identified by their parent, such as luxation of his shoulder and inability to go upstairs. There was history of intermittent luxations of the TMJ, weekly or biweekly.

On examination easy bruising, and ''cigarette- paper'' scars were found in the areas of trauma (Figure 1). He had habitual luxation of right and left shoulders, and joint hypermobility. We had observed hyperextension and hyperabduction of the fingers as well as a marked hypermobility of the elbows, feet and toes (Figure 2 and Figure 3). On cardio-pulmonary auscultation no evidence of mitral valve prolapse and cardio-pulmonary auscultation was normal. The peripheral pulse was also normal. In the examination of eye, the sclera was slightly blue (Figure 4). Bilateral clicking of the TMJ was felt at the end of

General consideration during treatment

	DO'S	DONT'S
Physical activity	Gently walk outside the home to socialize with peer group. Perform daily activities of living like dressing, eating etc. Develop individualized protocol of taking rest, and energy conservation.	Avoid straightening arms and knees in full swing, they will lock.
Exercise	Practice light resistance exercises for strengthening and stabilization. Learn appropriate exercise activities and precautions to strengthen muscles, protect joints, and prevent further damage.	Avoid high-impact exercises, like running, jumping, or physical contact sports, swimming.
Stretching	Be persistent and consistent in light resistance exercises. Stretching hamstrings can be performed.	Avoid most forms of stretching which further strains and loosens joints. Avoid yoga. Avoid heavy pulling, pushing and lifting, or jerky work or movement.
Skin care	Special attention should be given to skin care.	Avoid skin damage by using mild soaps, adhesive bandages.
Routine evaluation	Routine evaluation of the child's nutrition, growth, eyes, heart, skin, and joints is always needed.	Do not miss specialist's appointment.
Basic pain management	Application of heat, cold, electrical and ultrasound stimulation decreases pain during activity.	Avoid analgesics without prescription.
Surgical evaluation	Surgical evaluations to correct or prevent joint deformities should be considered as a last resort.	Avoid traumatic exposures which need surgical intervention.

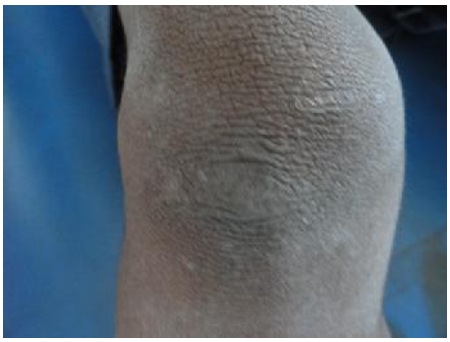

Figure 1. ''Cigarette- paper'' scars were found in the areas of trauma.

Figure 3. Hyperextension and hyperabduction of the fingers

Figure 2. Joint hyperelasticity

Figure 4. Blue Sclera

Figure 5. Intraoral photograph

Figure 6. Intraoral photograph of lower arch.

Figure 7. Intraoral photograph of upper arch.

Figure 8. Child with hyperelastic skin in orofacial region.

Figure 9. Child with hyperelastic skin in extrimity.

opening. The maximum opening was 54mm. Oral examination revealed a mixed dentition status. Oral examination revealed multiple carious teeth. Maxillary left central incisor was having ellis clss1 fracture due to fall. Gingival bleeding was also reported both spontaneously and after tooth brushing. (Figure 5, Figure 6 and Figure 7). Hyperelasticity of skin was also evident (Figure 8). Blood, electrolyte and liver function tests had exhibited within normal limit. Patient was too anxious so no radiographic evaluation was possible**.** So our case was diagonosed as Classical (Previously EDS Types I & II) type EDS.

Pedodontic consideration

A pediatric dental surgeon, pediatrician or dermatologist may be the first doctor to recognize EDS in a child patient. Management of EDS requires a team approach, including evaluation by a dermatologist, cardiologist, orthopedic surgeon and pediatric dental surgeon. EDS is considered to be a severe and painful chronic musculoskeletal disorder that may significantly reduce a person's effective quality of life, namely, recreation, home management, alertness, behavioral pattern, social relationship, movement, sleep and rest.[15] Pain is severe and common manifestation in EDS. The results showed that chronic pain in EDS is highly prevalent.[16] Acute disabilities due to EDS results in fractures, dislocations, subcutaneous bleeding or bruising after falls.[17] These may be happened in orodental structures also. Greater than three-quarters of EDS patients may be suffered from severe fatigue. Severely fatigued patients are more impaired than nonseverely fatigued and may have higher level of psychological distress often need counseling during dental management. Knowledge on behavior management helps a pododontost to treat such kind of cases in an easier manner. It is very unfortunate that there is no cure for EDS till date. Nonsteroidal anti-inflammatory agents may be prescribed for symptomatic relief of TMJ pain as well as dental pain. Preventative dental care is a part and parcel of treatment of EDS patients. Due to defective collagen synthesis EDS causes poor wound healing. So minor trauma in orodental region causes poor healing and small TMJ dislocations may be manifested in a greater dimension. Protective mouth guards may be useful in these cases. As a preventive measure child is advised to limit mandibular movements to avoid subluxation or luxation. If The child is suffering from of mitral valve prolapse prophylactic antibiotics are indicated for oral surgical procedures to prevent sub acute bacterial endocarditics.[18] Short duration dental appointment is always beneficial. Inferior alveolar nerve blocks should be given with optimum care to avoid causing hematoma.[18],[19] Finer gauze disposable optimally sterile needle is helpful. The buccal mucosa is vulnerable to traumatic injury from orthodontic appliances Forces in orthodontic management should be applied lighter than usual. The teeth may move rapidly with well-controlled forces, but the relapse is unavoidable, so retention is necessary for a longer period of time.[20] In children maxillofacial surgery should be avoided. If performed at all intra oral wounds after extraction should be covered with acrylic cover plate.[18] Surgery of the TM is only considered as a last resort to correct luxation or subluxation. There is no definite recommendation for general or local anesthesia in EDS child. Previous studies expressed reduced or no effects of local anesthetics in some patients.[21] EMLA cream can be used in some simple cases. General anesthesia can be performed with volatile anesthetics, nitrous oxide or total intravenous anesthetics. For preventive management of dental caries 1.23%APF /2% sodium fluoride solution application can be beneficial to EDS children. Conservative management is also essential in these cases. Selection of soft tooth brush and proper oral hygiene maintenance instruction and regular follow up are the important treatment protocols for EDS children.

Conclusions

Stomatognathic examination can be helpful in establishing diagnosis of EDS. Intraoral tissue fragility, functional impairment in daily life, reduced physical activity, psychological distress, recurrent TMJ dislocation are the challenges we may face during the dental treatment of an EDS child which may affect prognosis of dental treatment in an active and passive manner. Vascular EDS needs a special consideration because it may lead to premature death due to spontaneous arterial rupture. A wholesome attention, thorough treatment planning, multidisciplinary approach and skillful pedodontic hand is very much essential in treating these kind of children.

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