Oral manifestations and dental considerations of hereditary haemorrhagic telangiectasia in paediatric population — a systematic review

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Симптомы в полости рта и стоматологические аспекты наследственной геморрагической телеангиоэктазии у детей — систематический анализ

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INTRODUCTION:

One in 5000-8000 affected individuals in the population in general, is due to autosomal dominant hereditary disorder such as Osler-Weber-Rendu syndrome like Hereditary Haemorrhagic Telangiectasia, (HHT). Malformed vascular networks and endothelial cells are due to mutations in genes such as endoglin or ACVR1 which might affect the TGF-beta superfamily. Right-to left shunts are caused by lack of capillaries intervening the veins and arteries in case of arteriovenous malformations in HHT. Severity of HHT increases with the age. After removal of the pressure, refill occurs in which, might come before pressure blanching in relation to Telangiectasias in oral cavity and nasal cavity. Minimal trauma may result in tearing of lesions which are usually seen in superficially in relation to mucosal surface. Repeated bleeding from nasal areas is most common clinical presentation of HHT[1,2,3,4,5]. Arteriovenous malformations may occur in lungs, hepatic region or cerebral system such as PAVM,and CAVM. Bacteraemia might result due to performance of dental procedures such as removal of tooth, cleaning of teeth and root canal therapy. Presence of anaerobic bacteria is seen more than the aerobic bacteria. There is a relationship between the per iodontal abscess and brain abscess. Curaçao criteria consists of repeated bleeding from nasal region, occurrence of Telangiectasias in relation to oral cavity, labial mucosa, fingers, nasal mucosa and mucosa of gastro intestinal tract, AVM in brain, spinal cord, liver and the lungs and the first-degree relative with HHT[6,7,8,9,10].

The main objective of this study was to determine the oral manifestations of Hereditary Haemorrhagic Telangiectasia in paediatric patients and fill the gap between the paediatrics and dentistry.

MATERIALS AND METHODS:

This systematic review was followed as per the PRISMA guidelines.

ELIGIBILITY CRITERIA:

Research articles conducted in English such as randomised controlled trials, cohort trials, case control trials, cross sectional studies, opinion articles and case reports pertaining to paediatric population in relation to HHT were considered.

SEARCH STRATEGY:

Detailed and relevant documents search was conducted in the following databases such as MEDLINE. Duplicate records were removed to improve the accuracy. All the 5 articles were identified and screened. The search strategy included names such as Rendu, Osler, Weber syndrome, Hereditary, Haemorrhagic, Telangiectasia, paediatric population, oral and dental. The selected articles were read fully and summarised in following text.

RESULTS:

The findings of this review were based on 6 full text articles pertaining to oral manifestations of Hereditary Haemorrhagic Telangiectesia in paediatric population only. The article selection includes reviews, narrative reviews and original research. Dental professions are the first persons

Table 1. **Genes involved in HHT**
Таблица 1. **Гены, участвующие в развитии НГТ**

Gene	Chromosome Locus	Protein
ACVRL1	12q13.13	Serine/threonine-protein kinase receptor R3
ENG	9q34.11	Endoglobin
SMAD4	18q21.2	Mothers against decapentaplegic homolog 4

to come across to identify the oral manifestations of Hereditary Haemorrhagic Telangiectesia in paediatric population. The most commonly affected sites are the oral mucosa, gingiva, palate, tongue, nasal mucosa, etc.

General practitioner:

There is a link between PAVM and brain abscess. Bacteraemia can be formed even though regular tooth brushing is done. Dental procedures for the patients with PAVM and HHT should get the written consent for receiving the antibiotic prophylaxis prior to the dental treatments[11,12,13,14,15].

Oral Medicine:

Malformed capillary beds which blanch upon application of pressure are known as Telangiectasias. Specifically, Telangiectasias do not need any treatments.

Oral and maxillofacial surgery:

If we need any medical intervention, it can be managed by convention method, laser and electron usage[16,17,18,19,20].

Endodontics:

Endodontic therapy can be done in paediatric patients with HHT after getting proper informed consent.

Periodontology:

The periodontal prophylaxis includes oral hygienic instructions, rules and regulations, plaque control and scaling[21,22,23,24,25].

In summary, this systematic review highlights the gaps of knowledge in dental and oral considerations

Table 2. **Methods of treatment of hereditary hemorrhagic telangiectasia**
Таблица 2. **Методы лечения наследственной геморрагической телеангиоэктазии**

Author	Year	Article type	Main Field	No. patients	Treatment modalities/ Clinical relevance
Meir Mei-Zahav et al	2006	Cross-sectional study	OM	14	Cauterization and laser ablation
Jamie Mc Donald and David Stevenson	2006	Review	OM	-	Humidification, topical moisturizing therapy, haemostatic products, antifibrinolytic therapy, ablation therapy, systemic antiangiogenic agents, septodermoplasty, and nasal closure.
Cesare Danesino et al	2023	Review	OM	-	Argon plasma coagulation Embolization, Stereotactic radio surgery, and surgery.
Priya Verma	2022	Review	OM	-	Teeth cleaning, tooth extraction, orthodontic treatment, endodontics, clotting factor replacement, fluoride supplements, oral hygiene instructions, pit and fissure sealants, twice daily tooth brushing, mouth wash, dietary advice, haematologist opinion, prophylactic antibiotic therapy, and anti fibrinolytics
Vanishree Halasagundhi Shivakumar et al	2022	Review	OM	-	Tooth extraction, teeth cleaning, fluoride supplements, oral hygiene instructions, tooth brushing twice daily, mouth wash, endodontics, clotting factor replacement, dietery advice, prophylactic antibiotic therapy, haematologist opinion and anti fibrinolytic agent.
Ennio Bramanti et al	2014	Cross-sectional study	OM	116	Correction of coeliac disease, and paediatric dental treatment procedures.

of paediatric patients with Hereditary Haemorrhagic Telangiectasia and emphasises the importance of more studies in the particular field which includes case reports, original researches, etc. This may act as a reliable evidence and guide in clinical decision-making[26,27,28,29,30].

CONCLUSION:

This systematic review has the limitation in the research mainly in the field of paediatric oral and maxillofacial manifestations in relation to Hereditary Haemorrhagic Telangiectasia. This article might impart consciousness among the dental professionals to identify the disease since they are the first line of people who come across the condition. Future research relies on the oral and maxillofacial pathology mainly with HHT focusing on the paediatric, emphasising on the antibiotic prophylaxis, dental treatment procedures to be done and prognosis of the patients for the long-term follow-up.

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