Evelyn González[1]
Gianluca Sottilotta[2]

[1] Titicos Foundation, USA
[2] Hemophilia Center, Great Metropolitan Hospital of Reggio Calabria, Italy

ONLINE JOURNAL OF HEMATOLOGY & MEDICINE

Editor: G. Sottilotta
Editor in chief : D. Greco Malara
e-mail: ojhm@hemonline.it
https://www.hemonline.it

Original Manuscript

Dental Interventions in Hereditary Vascular Diseases: Minimizing Bleeding Risk

Definition and Clinical Relevance

Hereditary vasculopathies are disorders affecting the structure or function of blood vessels, especially capillaries, arterioles, and venules. Although they do not directly alter platelet count or coagulation factor levels, structural vessel fragility can lead to mucosal bleeding, spontaneous hematomas, and poor wound healing. Diagnosis is usually clinical and may be confirmed via genetic testing or skin biopsy.

Main Hereditary Vascular Disorders Relevant to Dentistry

1) *Ehlers-Danlos Syndrome (Classical and Vascular Types)*

- Connective tissue disorder affecting collagen types III and V
- Vascular type is the most severe: arterial rupture risk, deep hematomas, and uncontrolled bleeding
- Oral signs: fragile gingiva, gingival recession, bleeding during gentle brushing
- High-risk for oral surgery and deep nerve blocks
- Requires interdisciplinary management with hematology and clinical genetics

2. *Hereditary Hemorrhagic Telangiectasia (HHT / Rendu-Osler-Weber Syndrome)*

- Arteriovenous malformations on mucosa, skin, and internal organs
- Recurrent epistaxis, GI bleeding, oral telangiectasias (tongue, palate, lips)
- Chronic bleeding may cause iron-deficiency anemia
- High risk of mucosal bleeding during dental procedures
- Preoperative assessment with hematology is required; antifibrinolytics may be needed

3. *Simple (Constitutional) Purpura*

- Idiopathic capillary fragility, more common in young women
- Hematological values are normal
- Presents with spontaneous bruising and oral petechiae after minimal trauma
- No systemic therapy needed, but preventive dental care and antifibrinolytic rinses may be indicated in cases of prior prolonged bleeding

4. *Marfan Syndrome*

- Connective tissue disorder affecting elastic fibers
- Associated with aortic root dilation, valve prolapse, and cardiac risk
- In dentistry: evaluate cardiac status and anesthetic risk prior to any surgical procedure
- Less hemorrhagic but relevant for surgical safety)
- Mandatory cardiology clearance for patients with known cardiac involvement

Tests the Dentist May Request (Based on Clinical Suspicion):

- Complete blood count (usually normal) • Bleeding time (if bleeding is unexplained or prolonged)
- Genetic evaluation (conducted by a specialist) • Referral to hematology, internal medicine, or clinical genetics

Common Oral Findings:

- Telangiectasias on tongue, gingiva, and palate
- Submucosal hematomas
- Spontaneous gingival bleeding or prolonged bleeding after prophylaxis
- In some cases, poor postoperative healing or wound dehiscence

Recommended Dental Management:

1. Pretreatment Evaluation:
- Comprehensive history including family history and signs of connective tissue disorders
- Referral to hematology and genetics for diagnostic confirmation if not previously diagnosed

2. Non-Invasive Procedures:
- Generally safe with caution to avoid soft tissue trauma
- Antifibrinolytic rinses (e.g., tranexamic acid) recommended if bleeding history is present

3. Invasive Procedures (extractions, periodontal surgery):
- Only under medical clearance and appropriate hemostatic coverage
- Atraumatic surgical technique with firm sutures, prolonged pressure, and local antifibrinolytics
- Hospital-based care may be indicated for severe cases

4. Anesthesia Considerations:
- Infiltrative anesthesia is preferred
- Avoid deep nerve blocks if vascular fragility (e.g., Ehlers-Danlos vascular type) is suspected

Referral to Specialists

Any patient with suspected hereditary vasculopathy must be referred to hematology and/or clinical genetics for full evaluation, definitive diagnosis, and surgical risk assessment prior to dental intervention.

References

1. da Silva Santos PS, Fernandes KS, Magalhães MH. Osler-Weber-Rendu syndrome--dental implications. J Can Dent Assoc. 2009 Sep;75(7):527-30.
2. Tetsch NJ, Köppe J, Daume L, et al. Oral health and oral health-related quality of life in people with Marfan syndrome: a cross-sectional study. BMC Oral Health. 2025 Jun 5;25(1):925.
3. Amberger A, Pertoll J, Traunfellner P, et al. Degradation of collagen I by activated C1s in periodontal Ehlers-Danlos Syndrome. Front Immunol. 2023 Mar 7;14:1157421.

Conflicts of Interest: The authors stated that they had no interests which might be perceived as posing a conflict or bias.

Authors contributions: EG and GS contributed to the preparation of this manuscript.

Corresponding author: Dr. Evelyn González. Titicos Foundation (USA). E-mail: evealmamia@gmail.com