

Plasminogen Deficiency, Type 1: For Dentists



Overview of Plasminogen Deficiency (PLGD)

- PLGD Type 1: a quantitative protein deficiency, with decreased plasminogen activity and decreased antigen
 - Due to a genetic mutation in the gene for plasminogen; > 50 different mutations have been identified; Autosomal Recessive inheritance
 - Most common presenting symptom is ligneous conjunctivitis, but multi-organ, systemic disease that can be life-threatening
- PLGD Type 2: reduced functional activity of plasminogen, but normal antigen levels; patients are asymptomatic

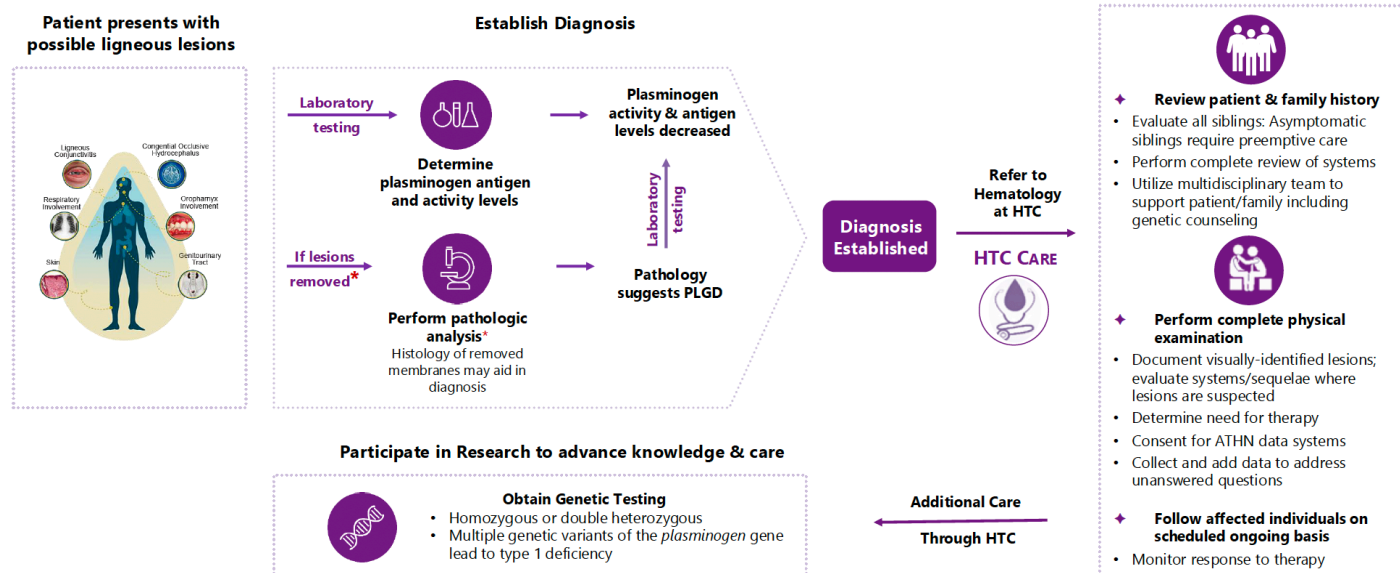
	Normal	PLGD Type 1	PLGD Type 2
Plasminogen Activity	70-130%	Decreased	Decreased
Plasminogen Antigen	6-25 mg/dL	Decreased	Normal
For Patients: My Plasminogen Activity			

Diagnosis

- Complicated by heterogeneous symptoms; symptoms can wax and wane
- Mucosal surfaces of the eyes, ears, nose, gums, airways, lungs, GI tract, kidneys, GU tract, CNS, and skin can all be affected
- Initial point of medical contact therefore includes many disciplines


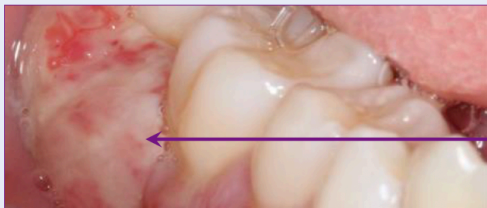
Treatment

- Ryplazim (plasminogen, human-tvmh) given by IV infusion leads to resolution of lesions
- Surgical removal of lesions, though initially helpful, leads to accelerated regrowth
- Referral to a Hemophilia Treatment Center (HTC) to serve as medical home, and:
 - Educate on product use
 - Ongoing symptom monitoring, outcomes, safety
 - Administer doses, determine dosing schedule
 - Teach home infusion



*Lesion removal prior to diagnosis not recommended

Dental Specific Diagnostic Findings

INITIAL PRESENTATION		ADVANCED DISEASE
<ul style="list-style-type: none"> ◆ Gingivitis ◆ Gingival pain ◆ Gingival bleeding ◆ Ligneous lesions ◆ Non-tender plaques 	Untreated	<ul style="list-style-type: none"> ◆ Ligneous lesions ◆ Periodontitis ◆ Gingival enlargement/hypertrophy ◆ Gingival recession ◆ Loose teeth ◆ Avulsed teeth ◆ Edentulism ◆ Exposed nerve with pain ◆ Alveolar bone loss ◆ Exophytic ulcerative soft tissue
 <p>Ligneous plaques & gingivitis</p>		 <p>Exophytic ulcerative soft tissue mass</p>
Courtesy of IHTC		MacPherson et al. 2020. Ref. 2. Used with permission

Dental Specific Treatment Considerations

- Ligneous gingivitis / periodontitis is the second most common manifestation of PLGD, with 34-43% of patients exhibiting lesions
- New or suspected diagnosis:
 - Obtain diagnostic blood test (plasminogen activity level) or refer to hematologist to order
 - If confirmed, refer to HTC to establish care and for full work up; work closely with HTC moving forward
 - Patients may have more than one system affected at presentation or occurring over time
- Confirmed diagnosis:
 - Coordinate clinical care with existing care team at HTC
 - Send clinical notes and photos
- Maintenance procedures:
 - Gentle dental cleaning with limited manipulation of the gum tissue (try to avoid deep cleaning or scaling)
 - Routine cleaning by dental hygienist coordinated with HTC
 - Gentle flossing and soft toothbrush
 - Non-alcoholic dental rinses
 - Always refer to the HTC before deep cleaning, extractions, restorations, crowns, or any procedures that require local anesthetic to coordinate administration of IV plasminogen, if recommended by the hematologist
 - Be suspicious of common diagnoses as they may be PLGD-related
 - Painless white or pink gingival enlargement
 - Gingival overgrowth with ulceration

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