

# 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

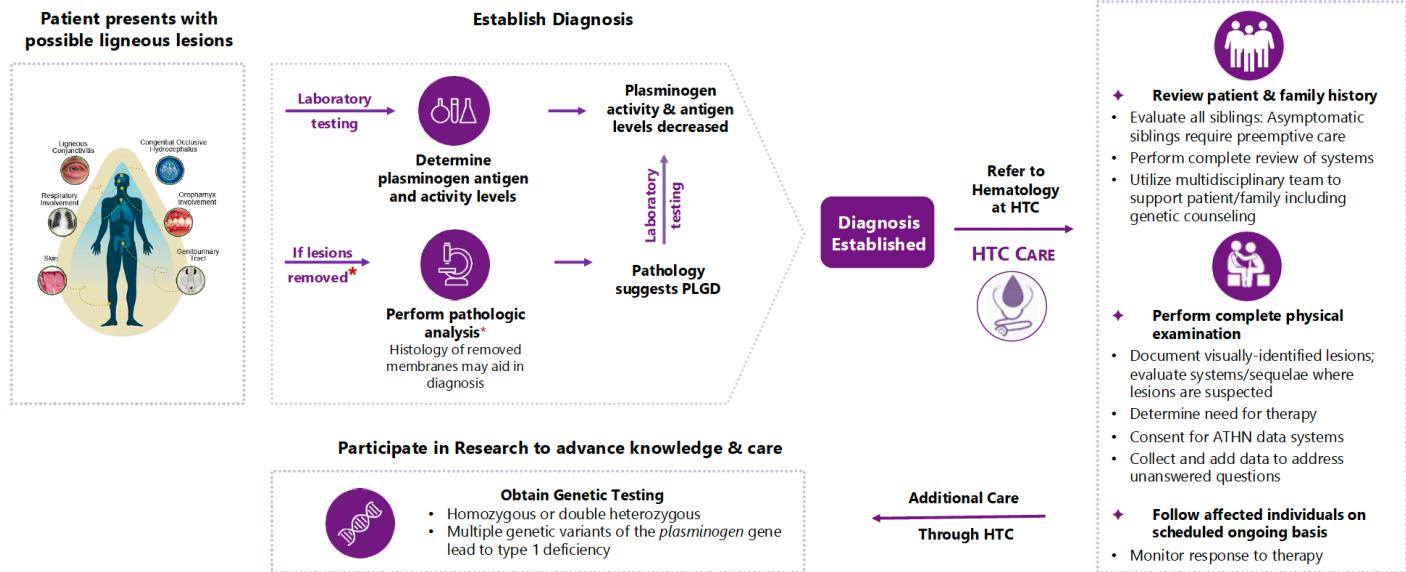
	Normal	PLGD Type 1	PLGD Type 2
Plasminogen Activity	70-130%	Decreased	Decreased
Plasminogen Antigen	6-25 mg/dL	Decreased	Normal
For Patients: My Plasminogen Activity			

## 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



## Dental Specific Diagnostic Findings

INITIAL PRESENTATION	ADVANCED DISEASE
<p>◆ Gingivitis ◆ Gingival pain ◆ Gingival bleeding ◆ Ligneous lesions ◆ Non-tender plaques</p> <p>Untreated</p>  <p>Ligneous plaques &amp; gingivitis</p>	<p>◆ 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>  <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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