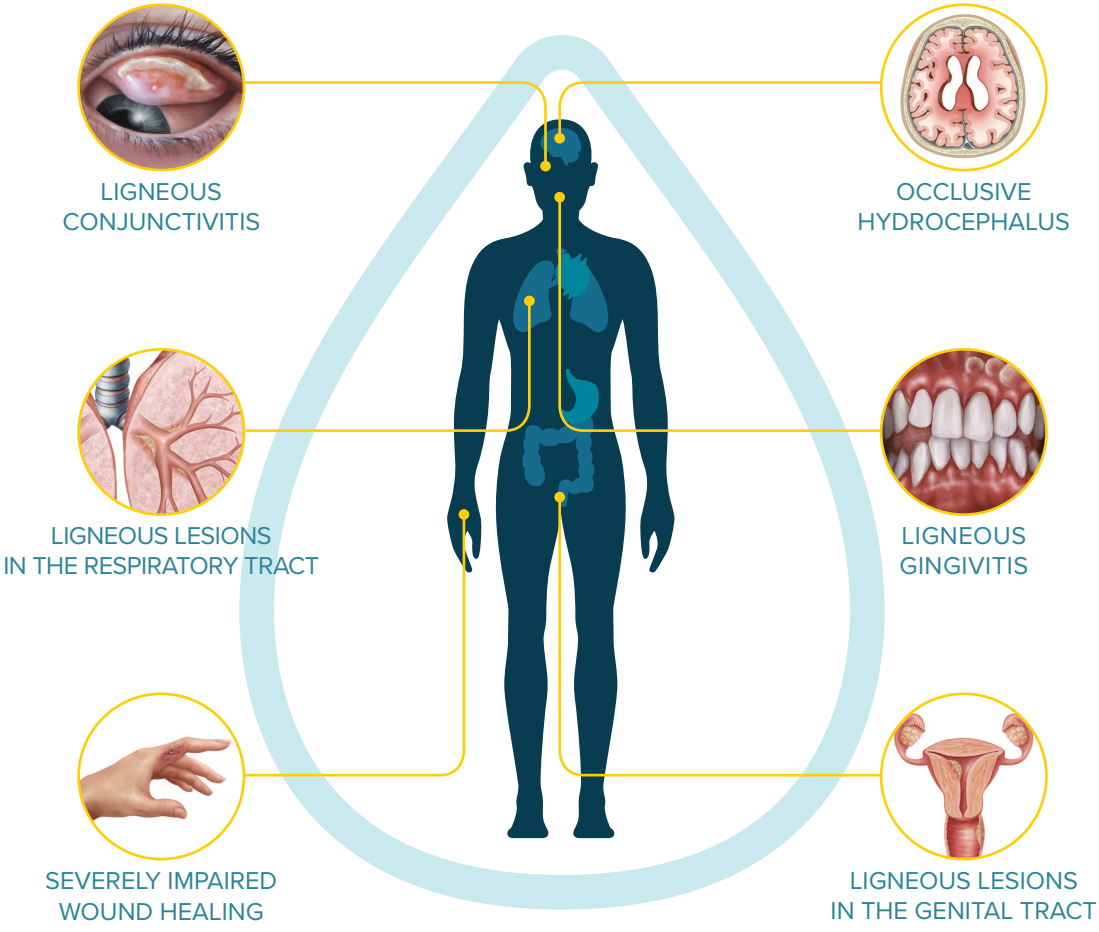
PLG|D CONNECTIONS

Congenital Plasminogen Deficiency (PLGD)

Patients are counting on you to recognize it

Congenital Plasminogen Deficiency (PLGD) is an ultra-rare genetic disease that can have devastating effects on multiple organ systems throughout the body.

Patients with PLGD may present with any of the following symptoms:



Reduced plasminogen activity can result in fibrinous lesions on a range of mucosal membranes throughout the body.

Test for PLGD if you recognize or suspect any of these clinical symptoms.

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Diagnosis Checklist

OBTAIN PLASMINOGEN ANTIGEN AND ACTIVITY LEVELS WITH ENZYMATIC BLOOD TESTS

- Plasminogen is an acute phase reactant; more than 1 assessment may be required.
- Levels may be affected due to anabolic steroid use, hypothyroidism, and hormonal contraceptives.

Form	Assay	Normal Levels
Activity	Chromogenic or fluorometric*	70-130%[1]
Antigen	Immunologic	6-25 mg/dL[1]

** Measurement of plasmin levels, following activation of plasminogen using streptokinase or urokinase*

- No specific levels have been identified that define or directly correlate with symptom severity. Activity levels <45% are thought to indicate deficiency.
- A comprehensive review of 74 patients with severe PLGD reported that functional plasminogen activity ranged from <1% to 51%[2].

CONDUCT A COMPLETE CLINCAL ASSESSMENT

Evaluate and document ligneous lesions. Histology of removed membranes can be helpful if diagnosis is unclear.

Request a genetic test as it will provide homozygous or heterozygous mutation status

- Multiple genetic mutations of the PLG gene are known to cause type I deficiency. K19E mutation is most prevalent (reported in 34% of identified patients)[3].

Confirm PLGD diagnosis with blood lab testing for plasminogen activity and antigen levels.

Evaluate family history as it may support diagnosis.

Recommend genetic counselling for patients.

Evaluate asymptomatic siblings as the disease can manifest at any age.

As a hematologic disease with multi-organ involvement and significant clinical sequelae, hematologists may be coordinators of multi-disciplinary care teams for patients with PLGD.

PLGD = Congenital Plasminogen Deficiency

References:
1. Mehta R, Shapiro AD. Plasminogen deficiency laboratory evaluation. Rare Coagulation Disorders.www.rarecoagulationdisorders.org/diseases/plasminogen-deficiency/laboratory-evaluation Accessed October 30, 2015 **2.** Schuster V, Hugle B, Tefs K. Plasminogen deficiency. J Thromb Haemost 2007;5:2315-22
3. Tefs K, Gueorguieva M, Klammt J, et al. Molecular and clinical spectrum of type I plasminogen deficiency: a series of 50 patients. Blood. 2006;108(9):3021-3026