

Leukocyte Adhesion Deficiency

Leukocyte adhesion deficiency (LAD) is a rare disorder characterized by recurrent bacterial infections caused by defects in neutrophil adhesion and delayed separation of the umbilicus. The molecular defect in patients with LAD is a deficiency of the beta 2 integrin subunit, called CD18. This subunit is responsible for making the protein LFA1 integrin, which allows neutrophils to make their way out of the bloodstream by binding to the Ig family receptor ICAM on the surface of endothelial cells in the infected areas of the body. The defect in neutrophil adhesion results in poor chemotaxis and phagocytosis, as well as elevated levels of neutrophils in the blood due to the inability to extravasate into tissue. Individuals with this deficiency suffer from bacterial infections beginning in the neonatal period that are often life-threatening. It is important to note that the infections are non-pyogenic, because neutrophils cannot migrate to the site of infection.



PLAY PICMONIC

Pathophysiology

Autosomal Recessive

Recessive-chocolate

Leukocyte Adhesion Deficiency is a rare disease inherited in an autosomal-recessive pattern.

Integrin CD18 Protein

CD (1) Wand (8) Ball

The molecular defect in patients with LAD is a deficiency of the beta 2 integrin subunit called CD18. This subunit is responsible for making the protein LFA1 integrin, which allows neutrophils to make their way out of the bloodstream.

Defect in LFA-1

Broken LooFA (1) Wand

The molecular defect in patients with LAD is a deficiency of the beta 2 integrin subunit called CD18. This subunit is responsible for making the protein LFA1 integrin, which allows neutrophils to make their way out of the bloodstream.

Attached to ICAM

Eye-camera

The LFA1 integrin allows neutrophils to make their way out of the bloodstream by binding to the Ig family receptor ICAM on the surface of endothelial cells in the infected areas of the body.

Signs and Symptoms

Neutrophilia

Nude-trojans

The defect in neutrophil adhesion results in poor chemotaxis and phagocytosis, as well as elevated levels of neutrophils in the blood due to the inability to extravasate into tissue.

Bacterial Infections

Bacteria-guy

The defect in neutrophil adhesion results in poor chemotaxis and phagocytosis, causing individuals with this deficiency to suffer from recurrent bacterial infections that are often life-threatening.

Non-Pyogenic

Nun Pus-pie

It is important to note that the infections are non-pyogenic because neutrophils cannot migrate to the site of infection.

Delayed Separation of Umbilical Cord

Unseparated Umbilical-cords

Delayed umbilical cord separation beyond the normal range of 3-45 days can raise suspicion of Leukocyte adhesion deficiency because this is common in infants with LAD.