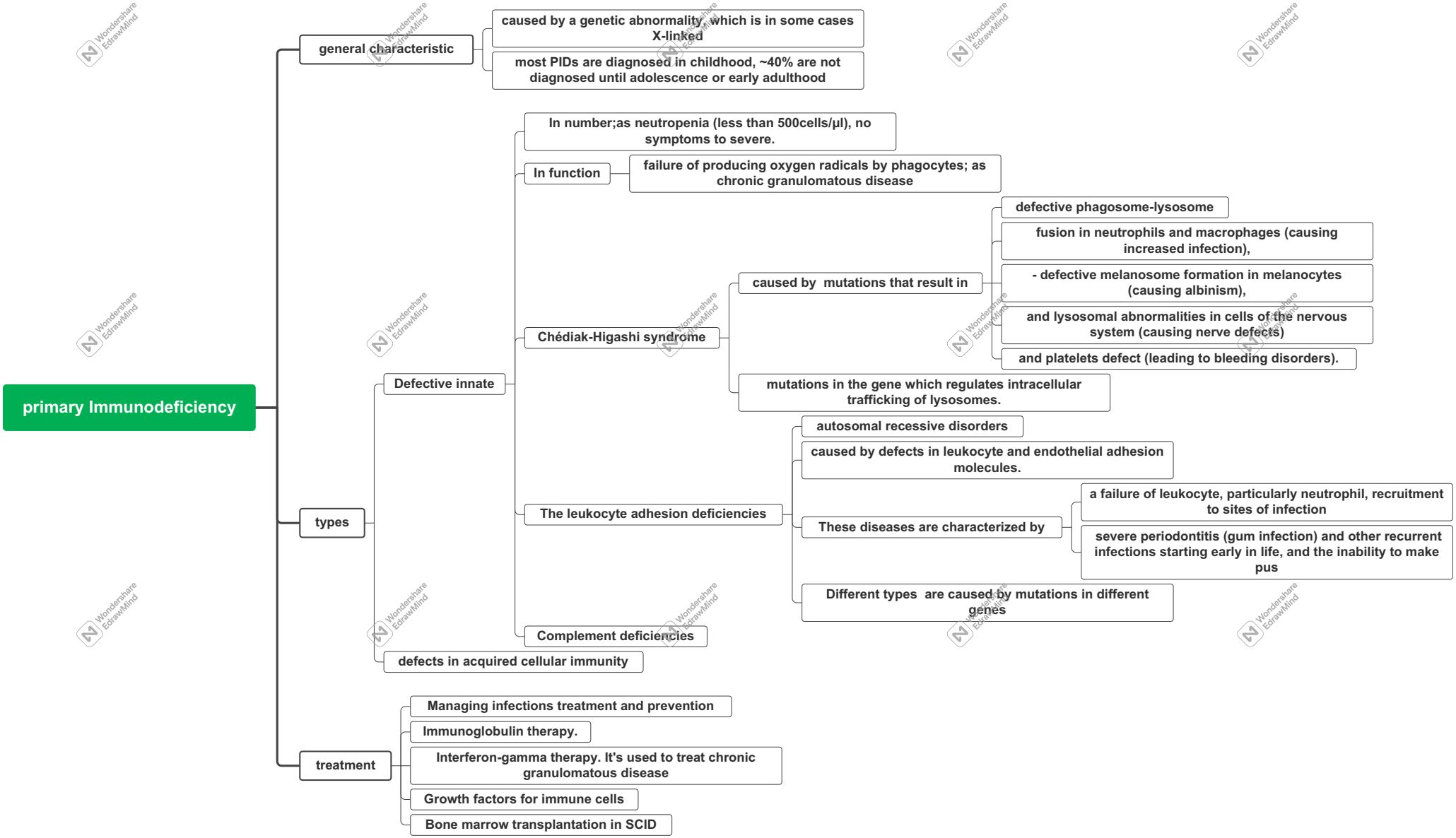
primary Immunodeficiency
general characteristic
caused by a genetic abnormality, which is in some cases X-linked
most PIDs are diagnosed in childhood, ~40% are not diagnosed until adolescence or early adulthood
types
Defective innate
In number;as neutropenia (less than 500cells/µl), no symptoms to severe.
In function
failure of producing oxygen radicals by phagocytes; as chronic granulomatous disease
Chédiak-Higashi syndrome
caused by mutations that result in
defective phagosome-lysosome
fusion in neutrophils and macrophages (causing increased infection),
- defective melanosome formation in melanocytes (causing albinism),
and lysosomal abnormalities in cells of the nervous system (causing nerve defects)
and platelets defect (leading to bleeding disorders).
mutations in the gene which regulates intracellular trafficking of lysosomes.
The leukocyte adhesion deficiencies
autosomal recessive disorders
caused by defects in leukocyte and endothelial adhesion molecules.
These diseases are characterized by
a failure of leukocyte, particularly neutrophil, recruitment to sites of infection
severe periodontitis (gum infection) and other recurrent infections starting early in life, and the inability to make pus
Different types are caused by mutations in different genes
Complement deficiencies
defects in acquired cellular immunity
treatment
Managing infections treatment and prevention
Immunoglobulin therapy.
Interferon-gamma therapy. It's used to treat chronic granulomatous disease
Growth factors for immune cells
Bone marrow transplantation in SCID