POLG Disease

Why in the News?

The son of Prince Robert of Luxembourg and Princess Julie of Nassau, Prince Frederik recently died from POLG disease.

- POLG disease is a genetic disorder that *depletes the body's cells of energy*.
- This in turn causes progressive multiple organ dysfunction and failure.
- It is a rare, incurable condition caused by inherited gene mutations.
- **Cause -** The condition affects mitochondria, the "powerhouse" of the cell which converts food into ATP, the primary energy our body uses to function.
- Mitochondria contain their own DNA which requires a specific enzyme to replicate, found in host cell's POLG and POLG2 genes.
- Mutations in these genes therefore impact the ability of mitochondria to replicate.
- **Symptoms -** Can start from early childhood to adulthood and can be mild to severe.
- The most frequent symptoms include muscle weakness, ophthalmoplegia (weakness of paralysis of the eye), epilepsy and liver failure.
- It could also affect a person's vision due to the involvement of affected brain structures.
- People with PolG likely end up bedridden and unable to function in daily activities.
- **Diagnosis** -It can be very difficult to diagnose because it affects a wide variety of organs and has many symptoms.
- **Treatment** - There is ***currently no*** treatment or cure for POLG.
- **Prevalence** – It affects 1 in 5,000 people.

References

1. The Hindu | POLG Disease
2. News 18 | What is POLG?