Juvenile Huntington's Disease (JHD)

Prelims: *Current events of national and international importance | Health*

Why in news?

First Juvenile Huntington's Disease brain donated to **National Institute of Mental Health and Neurosciences** *(NIMHANS), Banglore for research.*

- JHD – A ***rare inherited genetic disorder*** that causes progressive neuro-degenerative disorder that affects children and adolescents, causing progressive loss of movement control, thinking abilities, and emotions.

  Huntington's Disease – A genetic, adult-onset neurodegenerative disorder, but in rare cases, the disease begins ***before the age of 20*** *and is classified as* ***Juvenile HD****.*

- **Caused by –** A genetic change (mutation) of the ***HTT gene*** causes Huntington's disease. HTT gene makes a protein called huntingtin.
- The genes being passed on to a child by one of their parents, having the altered gene causes damage to certain areas of brain.
- Most often, children with juvenile HD inherit the genetic change repeat ***from their fathers***, although on occasion they inherit it from their mothers.
- **Symptoms –** Includes rigidity, stiffness, clumsiness, cognitive decline, and behavioral changes like irritability or mood swings, finding it difficult to control your muscles, eye movement, speech, and swallowing and the symptoms slowly get worse over several years.
- **Diagnosis –** Based on a combination of clinical symptoms, family history, and genetic testing, which identifies a ***larger-than-normal CAG (cytosine-adenine-guanine) repeat*** in the HTT gene.
  - The length of the CAG repeat expansion determines the age of onset - the longer the expansion, the earlier and more severe the disease.
- **Treatment –** Several types of treatment including Physical therapy or occupational therapy, Speech therapy, Counseling, Medications.
- Currently, this disease is ***not be cured***, but there are treatments to help manage the symptoms and stay comfortable and independent for as long

as possible.

References

1. The Hindu | 1st Juvenile Huntington's Disease brain donated
2. Cleveland Clinic | Juvenile Huntington Disease

