



## Juvenile Parkinsonism with Mental Retardation

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### Abstract

The investigations like complete blood picture, liver function test, renal function test, serum copper and ceruloplasmin, serum electrolyte levels, viral screening test were normal. No brain parenchymal pathology was detected in MRI scan. The patient is not a product of consanguineous marriage, the younger girl sibling was normal and no other member of the family juvenile Parkinson's is a form of Parkinson's disease that affects children and young people under the age of 20. Incidence of this type of Parkinson's is very rare. Its clinically and etiologically heterogeneous entity. Unlike the adults form secondary causes, hereditary and metabolic conditions are the predominant cause. 15yr old Male patient attended medicine OP with severe mental retardation, severe constipation, pill rolling movements. Patient's guardian gave History of delayed labor, delayed milestones, an episode of physiological jaundice, generalized tonic clonic seizures. On examination had masklike faces, mongoloid face with slanting eyes, big tongue, occasional resting tremors, MMSE score was 7/30 suggestive of severe mental retardation similar complaints. This is a rare case of juvenile parkinsonism with mental retardation

**Keywords:** Juvenile parkinsonism, mental retardation

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### Case history

A 15year old male patient resident of Sullurpet, studying 6<sup>th</sup> class presented to general

medicine OPD at Narayana Medical College, Nellore with chief complaints of repeated failure in class 5 and 6, inability to recall words and repeat phrases, difficulty in problem solving, following written commands, sentence formation and drawing geometrical figures since childhood. Irregular passage of stools since 6 years and abnormal movements of hands since 3 years.

In the Prenatal period regular antenatal checkups were done, no anomalies were detected during scans, no fever, iron folic acid supplementation were taken by mother. The labor was delayed, vaginal delivery at hospital and the baby cried immediately after birth. Regular postnatal followup, immunized

following universal immunization schedule. One episode of physiological jaundice and generalized tonic clonic seizures was noted. Delayed milestones.

Not a product of consanguineous marriage. No similar complaints in any other family members. The younger girl sibling is healthy. Normal sleep, appetite and bladder habits. Disturbed bowel habits i.e. severe constipation relived on taking medication. No smoking, alcohol or drug addictions. The patient had another attack of generalized tonic clonic seizures at the age of 3years and was on anti-epileptic medications for 2 years. No history of any drug allergies. No past surgical history.



**Figure 1:**

Parkinson's disease (PD) is the second most common neurodegenerative disease. Approximately 95% of cases of PD are idiopathic most likely caused by environmental factors and genetic susceptibility. [1] Unfortunately by the time of diagnosis most of the dopaminergic neurons in the substantia nigra are dead. Diagnosis of PD is based on classical motor symptoms including resting tremor, rigidity, bradykinesia and postural instability.[2] Juvenile Parkinson's is a form of Parkinson's disease that affects children and young people under the age of 20. Like other forms of Parkinson's, it is a neurological disorder that manifests itself as rigidity or stiffness, resting tremors and difficulties with initiating movements. There is also a range of non- motor symptom. [3] Under the age of 20 than for someone to be diagnosed in their 50s or 60s. [4]

The major symptoms of PD include rigidity, tremor, bradykinesia of the limbs, and postural instability. [5] These symptoms result primarily from a deficiency of dopamine caused by selective degeneration of dopaminergic neurons in two regions of the brain, the substantia nigra pars compacta and striatum.[6] Another pathological feature of this disease is the presence of inclusion bodies, called Lewy bodies, in those surviving neurons.[7]

On conducting general examination, the patient was noted to be conscious, coherent, cooperative, but not well oriented to time and place. Moderately built and nourished.[8] No pallor, icterus, cyanosis, clubbing, edema, lymphadenopathy, no skeletal deformities, no neurocutaneous markers like hypo or hyper pigmented areas, cauf-au-lait spots were noted.

*Vitals:* Pulse: 82 beats/min normal rhythm, rate, volume and character. Blood pressure was 110/80 mm of hg in sitting posture, no orthostatic hypotension. Respiratory rate was 16 breaths per min. Normal body temperature.

On clinical examination of the nervous system: Right handed, normal sleeping habits. Intellectual functions: Average.

**Appearance:** Mongoloid face, slanting eyes, big tongue. Voluntary and emotional facial

**Sensory System:** Superficial sensations like touch, pain, temperature are felt. movements are limited and slow and include an expressionless face (hypomimia), decreased eye blinking, and a blank stare (masked facies). No delusions or hallucinations. Not in disturbed emotional state. Orientation to time is not there [0/5 in MMSE Score] and places is impaired [2/5 in MMSE score]. Immediate, long-term and recent memory normal.

**General intelligence:** abstract thinking, reasoning, judgment, calculation is impaired. Motor system examination was done and The cranial nerve examination was normal. Bulk and nutrition of muscle was a good, no wasting or hypertrophy. Tone of muscles normal.

**Speech and language:** Naming is good, fluency, comprehension, repetition, reading and writing are poor. Speech is monotonous and of low volume (hypophonia).

S No:	Areas	RIGHT	LEFT
1.	SHOULDER	5/5	5/5
2.	ELBOW	5/5	5/5
3.	WRIST	5/5	5/5
4.	HAND	100%	100%
5.	HIP	5/5	5/5
6.	KNEE JOINT	5/5	5/5
7.	ANKLE	5/5	5/5

**Table 1:** powers

**Reflexes**

**Superficial reflexes:** corneal, conjunctival, abdominal, cremastic, reflex present. Bilateral plantar flexor reflex.

**Deep reflexes:** Biceps, triceps: 1+, knee: 2+, ankle: 0

**Coordination:** finger-nose, finger-nose-finger, knee-heel test normal. No rapid movements, no pendular knee jerk, no nystagmus, no dysarthria.

**Gait:** Near normal gait. No swing of hands while walking.

**Involuntary Movemnts:** Pill rolling movements, occasional resting tremors are seen.

Deep sensations like joint position, vibration, passive movements, deep pain (proprioception) felt. Tactile localization, two point discrimination, steriognosis, and graphasthesia present. No signs of meningeal irritation like neck stiffness. No peripheral nerves thickening. Higher function testing using MMSE was done and the patient scored 7/30 (very severe mental retardation). No symptoms of raised intracranial tension like headache or vomiting's, sudden loss consciousness, relapse of convulsions. No fever, spinal cord trauma, drugs, dog bite **OTHER system examination:**

CVS: S1, S2 heard, no murmur.

RS: bilateral air entry and no vesicular breath sounds.

Git: No hepatomegaly, no splenomegaly.

**Investigations:** Complete blood picture, liver function tests, complete urine examination, ultrasound abdomen, serum electrolyte levels, viral screening were normal. No abnormal findings were noted in slit lamp examination. Fundus examination was normal .NO KF rings. NO sunflower Cataract. X-Ray chest was normal. Imaging modalities like MRI was conducted to help in ruling out other disease, was suggestive of normal brain parenchyma.

**Discussion**

From the history, detailed examination and investigations the following important findings are to be considered: 15yr old Male patient, severe mental retardation, severe constipation, resting tremors, pill rolling movements, masklike faces, no swinging of hands while walking, mongoloid face with slanting eyes, big tongue, delayed labor, physiological jaundice, generalized tonic clonic seizures.

Infectious cause was ruled out as the complete blood picture was normal, no elevated ESR, negative viral screening tests, no travelling to any disease endemic areas. Metabolic defects like that Wilsons disease was ruled out as serum ceruloplasmin, serum copper levels were normal,

no KF rings, no sunflower cataract in eyes, Liver function tests were normal, ultrasounds abdomen was normal. Other metabolic conditions like diabetes, thyroid not present. No evidence suggestive of neoplastic origin. The juvenile form of Huntington's disease was also ruled out by the absence of an antecedent familial history, no features in the MRI scan. No complaints or features of toxic poisoning. The pill rolling tremors, mask like faces, resting tremors, presenting at the 15 years of age is suggestive of juvenile parkinsonism. As the MMSE score is 7/30 the patient is having severe mental retardation which may be due to hypoxia due to delayed labor.[9] The patient may develop Shy Dragers syndrome in future as the patient has a history of severe constipation though orthostatic hypotension has not developed yet.[11] Currently the patient is started on small dose of L-Dopa [250mg BD] along with Pacitin [50mg]. The reporting of such cases can lead to better awareness of such rare diseases, the incidence of which is on an increasing trend. Further genetic studies may throw a light on different etiopathogenesis and pave way for a better treatment of this disease. If the genetic studies are suggestive of hereditary role then proper genetic counseling and more research in this area will be encouraged, thus contributing to knowledge and betterment of the society.

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