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Case Report

A CASE REPORT ON FAHR DISEASE

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ABSTRACT
Fahr disease is also known as Idiopathic Basal Ganglia Calcification is a rare, genetically, dominant neurological disorder. This disease is characterized by abnormal deposits of calcium in areas of the brain that control the movement. The present study shows different CT Scans through which different calcifications are seen and analyzed.

Keywords: *Fahr Disease, Idiopathic Basal Ganglia, CT Scan*

INTRODUCTION
Fahr disease is a rare, genetically dominant, inherited neurological disorder characterized by abnormal deposits of calcium in areas of the bran that control movement.
Grossman and Yousen (2003) and Shenoy *et al.,* (2009) reported the neurological disorder in case of Fahr's disease. To understand the intensity of the disease different radiological Images are mentioned. Shafaq *et al.,* (2013) suggested that early diagnosis and treatment can reverse the calcification process leading to complete recovery of mental functions.
Hoque *et al.,* (2010) reported a case of Fahr's disease, who presented with complex partial seizure and behavioral abnormalities.
The case of seizure on screening is also mentioned. Ring and Sera-mastres (2002) suggested that Fahr's disease is differentiated from Fahr's Syndrome in which basal ganglia calcification is secondary to some other disorder, such as hypoparathyroidism. Patients with Fahr's disease often appear with movement disorders such as Parkinsonesin, Paresis and speech impairment.

CASES
An 80 year old female was admitted with vomiting, diarrhea, slurred speech, difficulty in standing and walking since three days, known case of hypertension since 10 to 15 years and bronchial asthama since 5 years. Non diabetic non tuberculous physical examination showed mild dehydration and signs of bilateral cerebellar involvement. She had long standing memory and behavioural changes with fatiguability. Different investigations like CT Scan (Brain), 2D echo, Blood urea and CUE were made and analyzed.

DISCUSSION
FAHR Disease
Idiopathic Basal Ganglia Calcification, also known as Fahr disease, is a rare, genetically dominant, inherited neurological disorder characterized by abnormal deposits of calcium in areas of the brain that control movement. Through the use of CT scans, calcifications are seen primarily in the basal ganglia and in other areas such as the cerebral cortex and cerebekum. These changes are shown in Figure 1 & Figure 2.
A locus at 14q has been suggested, but no gene has been identified. A second locus has been identified on chromosome 8 and a third has been reported on chromosome 2.
The most commonly affected region of the brain is the lenticular nucleus, the internal globus pallidus. Calcifications in the caudate, dentate nuclei, putamenand thalami, occasionally calcifications begin or predominate in regions outside the basal ganglia.
Calcification is progressive, since calcifications are generally more extensive in older individuals Histologically concentric calcium deposits within the walls of small and medium-sized arteries are present. Droplet calcifications can be observed along capillaries. These deposits may eventually lead to closure of the lumina of vessels.

Usually found in the third to fifth decade of life with clumsiness, fatigability, unsteady gait, slow or slurred speech, difficulty swallowing, involuntary movements or muscle cramping. Seizures are common, difficulty with concentration and memory to changes in personality and/or behavior.

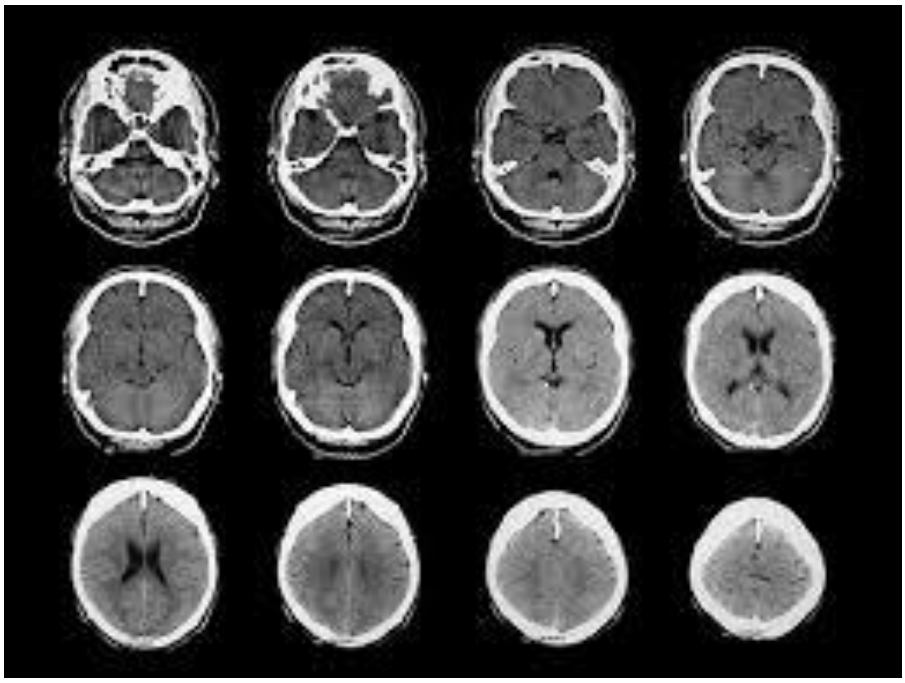

Figure 1: (CT Scan Brain - Normal)

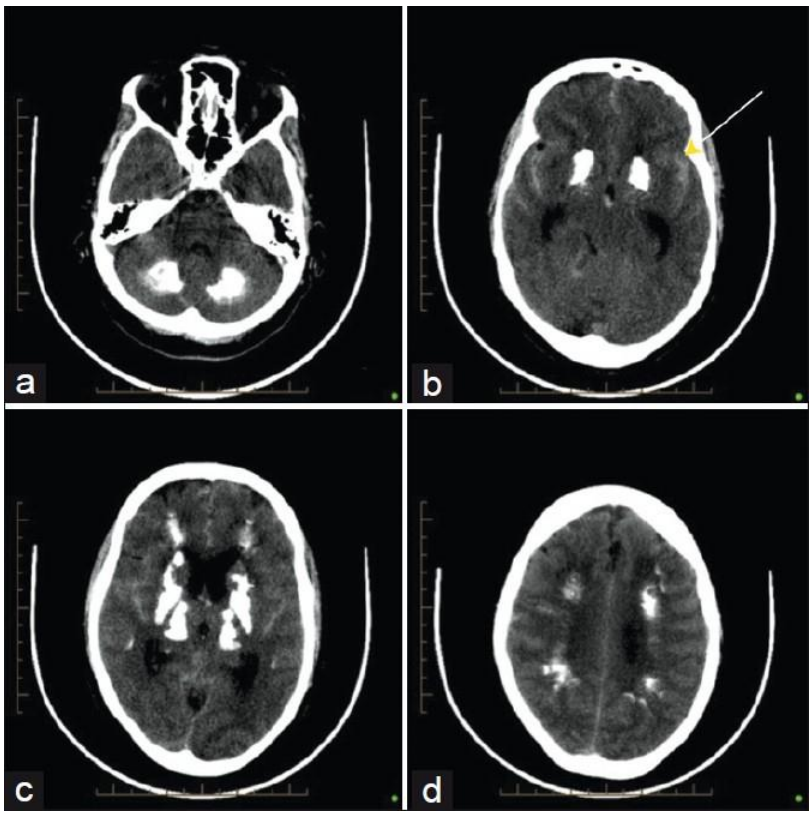

Figure 2: (CT Scan Brain - Fahr disease)

The diagnosis requires the following criteria be met:

- The presence of bilateral calcification of the basal ganglia
- The presence of progressive neurologic dysfunction.
- The absence of an alternative metabolic, infectious, toxic or traumatic cause
- A family history consistent with autosomal dominant inheritance.

The calcification is usually identified on CT scan but may be visible on plain films of the skull.

- CSF analysis – to exclude bacteria, viruses and parasites
- The Ellsworth Howard test (a 10-20 fold increase of urinary cyclic AMP excretion following stimulation with 200 micromoles of parathyroid hormone) may be worth doing also Serology for toxoplasmosis is also indicated.
- Brain CT scan is the preferred method of localizing and assessing the extent of cerebral classification.

Prognosis

The prognosis for any individual with Fahr's Syndrome is variable and hard to predict. There is no reliable correlation between age, extent of calcium deposits in the brain, and neurological deficit. Since the appearance of calcification is age-dependent, a CT scan could be negative in a gene carrier who is younger than the age of 55.

Progressive neurological deterioration generally results in disability and death.

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