

# Management of Mandibular Body Fracture of Patient Suffering From Huntington's Chorea : A Case Report

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## Abstract

Huntington's disease is a neurological disorder characterized by progressive dementia and worsening choreathetoid movements, conservative methods for mandibular fracture are not appropriate and open reduction under general anesthesia is primarily indicated. This report illustrates an unusual rigid fixation of body fracture of mandible which is minimally invasive and equally effective and requires short operating time in a patient of Huntington's chorea where patient was sedated as general anesthesia could not be given because of upper respiratory tract infection.

**Key words** : Chorea , sedation, splint, minimally invasive

## Introduction

Chorea, the Greek word for "dance," is used to describe the involuntary movements of the body especially of arms, legs and face . In Huntington's chorea, these movements tend to be spastic and ballistic, and they dramatically affect normal functioning.<sup>3,12</sup>

Huntington's chorea is a dominantly inherited disease<sup>10,13</sup> which is passed down through families by an autosomal dominant form of inheritance. It was first described by George Huntington in 1872<sup>2,6,12</sup>. Incidence is 5-10/100,000.

The disease often presents as "nervousness", and depression. Eventually progresses to include dementia and slowed eye movements. In juvenile HD, rigidity and parkinsonian tremor may be the primary manifestation. Generally, symptoms are first observed when the victim is in the '20s or '30s, and the degeneration slowly progresses, ending in the victim's death some 10 to 20 years later.<sup>2,12</sup>

The disease progresses with increasing chorea and loss of mental abilities. Eventually, abnormal movements and dementia become so severe that the person can no longer care for his or her self. Huntington's chorea is due to slow degeneration in the basal ganglia, which eventually leads to cell death in the brain and the decrease and increase of various neurotransmitters. The symptoms of the disease are caused by a significant reduction (volume and activity) of two principal neurotransmitters (naturally occurring chemicals in the brain) namely Acetylcholine and GABA, in turn affecting the activity of the neurotransmitter Dopamine, which becomes hyperactive. The disorder is partly characterized by an increase in the availability of dopamine, which can cause symptoms of chorea. Huntington's disease is a basal ganglia disease; the portions most severely affected are caudate and putamen<sup>1,12</sup>.

## Symptoms

Huntington's Disease is characterized by three types of symptoms: motor, psychiatric, and cognitive.

Motor dysfunction involves involuntary, uncontrolled movements. These can begin as minor twitches and tics, but eventually lead to a general loss of coordination and balance that make walking, speaking, and eating increasingly difficult.

Psychiatric symptoms are harder to diagnose, with the most common symptom being depression. This can include hostility and a loss of motivation. Many patients also become manic-depressive and experience strong mood swings. Delusions and hallucinations also sometimes result from the disease and can frequently cause the subject to suffer from feelings of paranoia<sup>10</sup>.

Cognitive impairment can be widespread, especially approaching the end stages of the disease. Simple tasks such as mathematics and routine chores can become a challenge as memory and decision-making become impaired. Often, the loss creates difficulty distinguishing these symptoms from those of a psychiatric nature.<sup>12</sup>

Huntington's disease is a dominantly inherited progressive autosomal disease that affects the basal ganglia. Symptoms appear later in life and manifest as progressive mental deterioration and involuntary choreiform movements. Patients with Huntington's disease develop a progressive but variable dementia. Dysphagia, the most significant related motor symptom, hinders nutrition intake and places the patient at risk for aspiration. The combination of involuntary choreoathetoid movements, depression, and apathy leads to cachexia.

## Case Report

A 50 year old frail woman reported in the emergency room with a history of fall from the bed and struck the side of bed . Patient was evaluated in the emergency room and diagnosed as having left body fracture of

mandible. Patient was examined by a neuropsychiatrist Patient's relatives gave a history of abnormal movements since ten years, loss of speech, hostile behavior with increasing dementia and difficulty in swallowing tone and gait in next 3-4 years. Presently her mini mental state

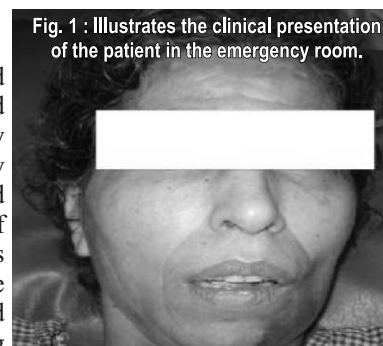


Fig. 1 : Illustrates the clinical presentation of the patient in the emergency room.

examination (MMSE) score was 10/30. Patients behavioral symptoms included wide emotional instability, hostility and on and off paranoid features. At the time of presentation patient was markedly agitated, hostile and uncooperative. Family history was positive for abnormal movements and behavioral problems in paternal uncle of the patient. There was no history of any psychoactive substance abuse, seizure disorder, paralysis, prolonged fever, meningitis, thyroid dysfunction, psychotropic medicine usage. A classical triad of symptoms (motor, psychiatric, cognitive) along with a positive family history strongly suggested a diagnosis of Huntington's Chorea. Patient was advised genetic typing to confirm the diagnosis but due to unavailability of the test in our and other medical centres the test was not done. OPG X-ray was taken to confirm the diagnosis of mandibular body fracture (figure 2). Radiographs both pre op and post op are of poor quality as patient's movements posed a hindrance for a good quality radiograph.

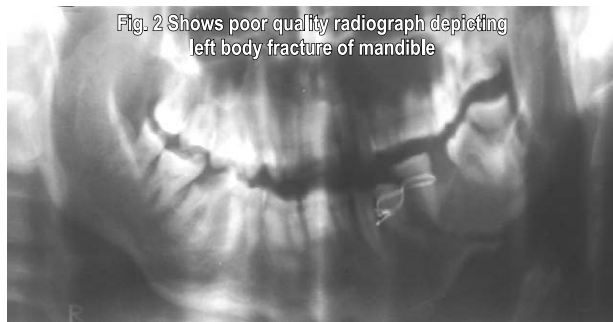


Fig. 2 Shows poor quality radiograph depicting left body fracture of mandible

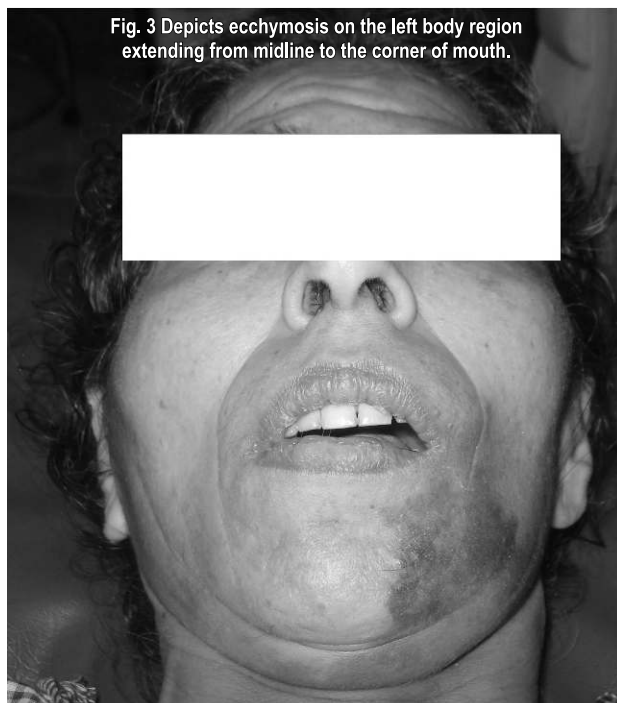


Fig. 3 Depicts ecchymosis on the left body region extending from midline to the corner of mouth.

On examination ecchymosis extraorally on left side of mandible (figure-3) and intraorally in left lower vestibule was present. In preprocedure assessment patient was found to be having widespread cognitive, significant motor impairment, hostile behavior and memory loss posed a

challenge for intermaxillary fixation so open reduction and internal fixation of her fracture was planned. Now concerns of anaesthetist came forth that is how to treat frail elderly women incapable of cooperation, suffering from malnourishment, having increased risk of aspiration<sup>7,8</sup>, could have exaggerated response to succinylcholine and thiopental<sup>4,7,9</sup> and also suffering from upper respiratory tract so general anaesthesia and intubation was avoided. It was clear that some modifications in the anesthetic technique would be required to successfully complete this case<sup>14</sup>. Patient was planned for short surgery under sedation and local anaesthetics. Fracture reduction was to be achieved by cap splint placement with circummandibular wiring. Impressions were made, cast prepared, osteotomy done at fracture site and occlusion achieved. A 20 gauge wire adapted around the buccal surface of lower teeth and another wire on lingual surface for better stability of fracture fragments. Acrylisation done, a cap splint was fabricated which was 4mm short of vestibule.

### Surgery

Patient was taken in operation theatre, surgical preparation of the patient done.

Patient was sedated with midazolam. cap splint was placed and checked for adaptation. Splint stabilized with circummandibular wiring (figure-4). The procedure was short and fracture reduction achieved with minimally invasive surgery



Fig. 4 Illustrates the stabilized mandibular occlusal splint

Immediate post op radiograph (figure-5) shows good approximation of fracture fragments and wires incorporated in the splint to maintain the strength and integrity of cap splint well adapted around the lower teeth. Soft diet was prescribed to the patient. Her recovery was uneventful and she was discharged the same day. Splint was removed two months later. post op X-ray showed good healing.

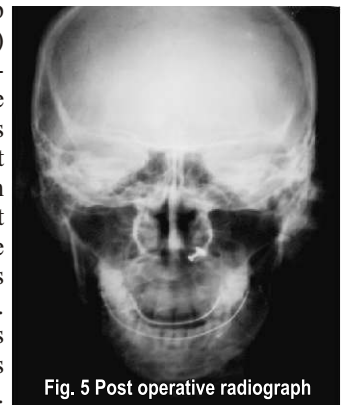


Fig. 5 Post operative radiograph

## Discussion

Jaw immobilization compounds preexisting speech difficulties and hamper post operative nutrition in patients with motor dysfunction such as huntington's chorea which represents a relative indication of open reduction of body of mandible<sup>1</sup>.

At present there is no cure for the disease so aim is to treat the symptoms and support and protect the patient. With open reduction patient's basic need of hygiene and feeding can be easily taken care of. But open reduction for these kind of patients can only be taken up under GA.

Anaesthetic triad consists of loss of consciousness (amnesia), analgesia, muscle relaxation. amnesia is achieved by midazolam as it is short acting, has rapid onset of action, rapid metabolism so no cumulative effects produced.

Analgesia is achieved by NSAIDs, opioids, nitrous oxide, vapour has been used with no relevant reports of side effects. Nitrous oxide requires an inducing agent like propofol or ketamine to provide anaesthesia<sup>11</sup> but the latter two drugs cause tremors so nitrous oxide was ruled out.

Muscle relaxation could not be achieved as this patient can be sensitive to non depolarizing muscle relaxants, depolarizing relaxants like succinylcholine have been used on such patients but reduced plasma cholinesterase activity with prolonged response to succinylcholine has been observed<sup>8,9,11</sup>. Use of sevoflurane and mivacurium for general anaesthesia in these patients have found to be safe<sup>7,14</sup> but availability of these drugs in India is still restricted. This patient was suffering from upper respiratory tract has a predisposition for laryngospasm and bronchospasm and will have difficulty in maintaining pulmonary ventilation So intubation of patient was ruled out and i/v anaesthesia (sedation with midazolam) was preferred for a short surgery to produce minimal cardiovascular and respiratory effects. Since the operating time was short and patient's uncontrolled movements and hostile behavior were a hindrance for placement of wires and achieving IMF and intraoral open reduction which is usually the recommended line of treatment for patients suffering from dementia<sup>1</sup>. Treatment options were zeroed in to placement of cap splint for fracture reduction which requires a preoperative fabrication time but a minimal operating time, the patient's discomfort remains minimal, the materials are cheap<sup>16</sup>. Since the movements of the patients were reduced once she was sedated, the procedure took twenty minutes to complete.

## Conclusion

All these collaborated into one proved to be a successful approach for the treatment of mandibular fracture of patient suffering from huntington's chorea. Short sedation evaded the complications of inducing agents and muscle relaxants and recovery of the patient was quick and eventful and she could be discharged the same day.

Cap splint was an effective method for reduction and fixation, minimally invasive with no hindrance in maintaining nutrition and hygiene and reasonably good post operative recovery.

Two month later patient had recovered fully from injury and Xray showed good bone healing.

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