Case Report

Satisfactory Improvement in Lifestyle of a Spastic Quadriplegic Cerebral Palsy Patient through Cranial Surgery

Md Faridul Islam Chowdhury

Abstract

Several treatment options may help improve daily functioning in children with cerebral palsy (CP). However, these treatments cannot prevent early death or enable them to lead independent life without support. CP is a progressive disease, and even in its milder form, conditions such as Parkinson's disease, epilepsy, and stroke may develop even after 25 years. A new era of curable treatment has emerged for CP, utilizing bilateral cranioplasty and duraplasty (Tanfarid Procedure). This case report describes a successful case where this procedure was employed. During the surgery, after the removal of cranial bones through craniotomy, the absence of dural pulsation was observed, indicating elevated intracranial pressure. After duraplasty, normal brain pulsation was observed, indicating that intracranial pressure had normalized. In patients with CP, premature closure of sutures and thickening of skull bones lead to a reduced intracranial volume, which hinders proper brain growth and eventually results in brain atrophy, widened Sylvian issures, and widened sulci accompanied by the loss of brain parenchymal tissue. These findings were also evident in the patient's brain CT scan. In this case, cranioplasty and duraplasty provided adequate space for the brain to grow. The treatment of CP patients typically involves a multidisciplinary approach, which can be expensive. However, the procedure utilized in this case is affordable for economically disadvantaged individuals in this country. While a second cranial surgery may be required in some cases, the expenses associated with it are significantly lower compared to a lifetime of physiotherapy and other supportive treatments. Additionally, the outcomes are very promising

Keywords: *Cerebral palsy, Birth asphyxia, Convulsion, Cranioplasty, Duraplasty.*

Date of received: *12.12.2022*

Date of acceptance: *15.05.2023*

DOI: https://doi.org/10.3329/kyamcj.v14i02.68533

KYAMC Journal. 2023; 14(02): 105-108.

Introduction

One of the most important causes of Cerebral Palsy (CP) is birth asphyxia which is due to prolonged labor with fetal distress, maternal distress, oligoamnios, Low birth weight, amniotic fluid aspiration or meconium aspiration, placental insufficiency, or maternal hypovolemic shock either due to spinal anesthesia or hemorrhagic shock which is not corrected in due time also the cause of CP[1] Hippocrates was the first who described the seizures and CP. However, these are not sufficient and well documented.[2]

Dr. Little began work on CP in late 1830s when he gave lectures on birth injuries. He is the first person to define oxygen deprivation at birth as a cause of brain injury and attempted to correlate oxygen loss at birth and brain damage which turn into CP. Sir William Osler, who wrote a book entitled “Cerebral Palsies of Children,[3] in 1887. Convulsion is the most common and devastating symptom of CP. Repeated convulsion causes cerebral hypoxia again and again and any anti-convulsive agents cannot prevent it. All types of epileptic seizures can be seen in patients with CP. Complex partial and secondary generalized ones are the most frequent seizure types.[4] These different types of convulsion every time lead to cerebral hypoxia which eventually cause further brain damage. Birth asphyxia leads to hyperostosis of the skull bones and facial bones. So the fontanelle and sutures close prematurely and actually these pathological processes lead to the abnormal shape of the head with raised intracranial pressure. Frontanalle and sutures are very important for the growth of all skull bones uniformly for taking normal intracranial volume . If the size and shape of the skull apparently looks normal in CP

01. Professor of Neurosurgery, Northern International Medical College, Dhanmondi, Dhaka, Bangladesh.

Corresponding author: Md Faridul Islam Chowdhury, Department of Neurosurgery, Northern International Medical College, Dhanmondi, Dhaka, Bangladesh. Cell Phone: +8801711481539, E-mail: dr_faridul@yahoo.com

patients, thickened skull bone scould be the cause to decrease intracranial volume. In this case, the head the patient looked like normal because of thick scalp. When the scalp from the skull was separated during operation, then the real size of the skull was appeared, smaller. Spastic cerebral palsy is the most common type of CP[5], such as spastic diplegia, spastic hemiplegia and spastic quadriplegia. In this case report, a successful treatment procedure of a such patient has been described.

Case Presentation

The parent's name Fatema, a one-year-old baby girl, came with complaints that her muscles are flabby and spastic, she was blind and underweight (4.8kg only). She was unable to grip, unable to raise her neck and head. She was also unable to turn herself from side to side, sit or crawling. Furthermore, she had difficulty in taking liquid food, was totally unable to take solid food, even she could not cry loudly, and had no interest in any toy or persons, she was admitted to Sheikh Fazilatunnessa Mujib Memorial KPJ Specialized Hospital & Nursing College (SFMMKPJSH&NC), Gazipur on 26 may 2022 and operated 30 may 2022. All her investigations were done including echocardiography. All were normal except body weight. No other congenital abnormality was found.

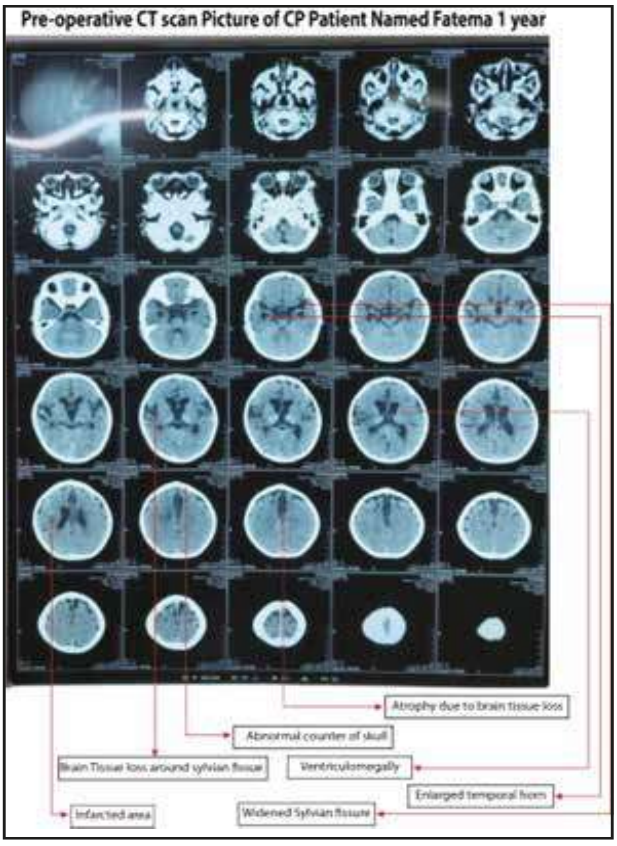


Figure-1

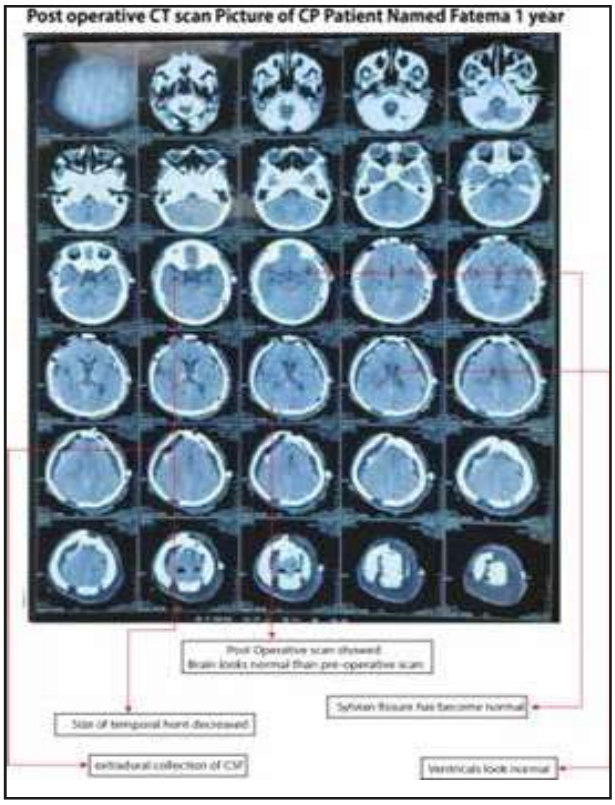


Figure-2

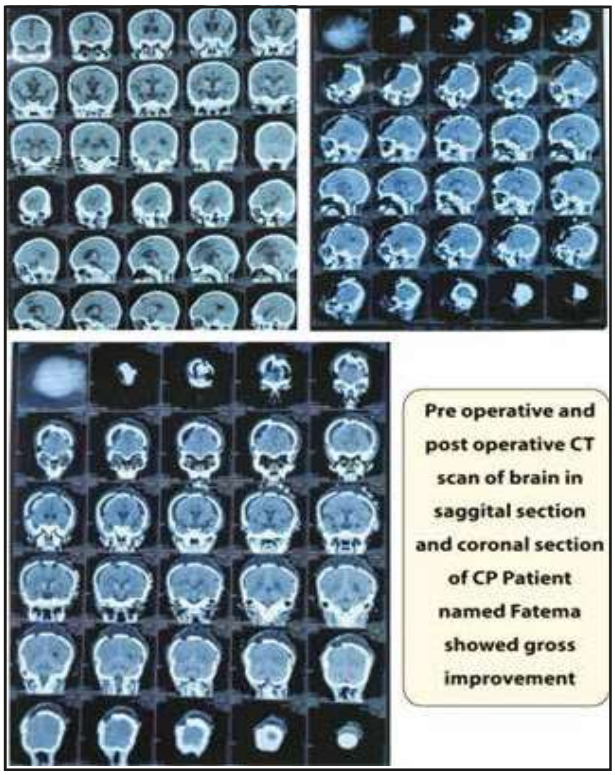
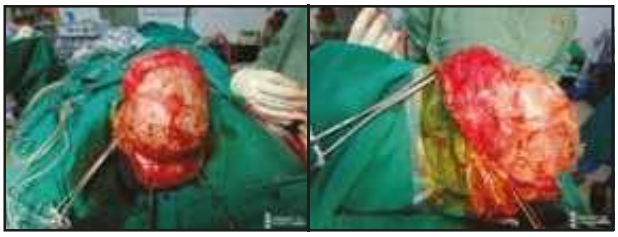


Figure-3: Per-operative picture of reconstructed skull, mid sagittal section infracted area. In coronal section fissure, ventricles and infarction all came near to normal Post-operative CT showed totally normal.

Invented surgical approach

Bilateral Cranioplasty and duraplasty and removal of skull base hyperostosis bone by drilling, removal of thickened temporal bone.



Per Operative Picture is temporal bone indrawned and skul strong like adult skull

The physical appearance of CP patient before operation, just after operation and 25 days after operation and four months after the operation

Discussion

Diagnosis of CP is easy as it is primarily based on clinical findings and clinical history. However, it is essentially needed to be confirmed by CT scan of brain. It can be diagnosed before the baby reach at one year of age. In the diagnosis of CP, MRI is also helpful in addition to a CT scan of the brain. Specially to detect periventricular hypo-density and early detection of hypo-intensity in other areas of the brain. Only MRI is not the ideal one, because skull bones cannot be visualized by it. CT scan help to detect widened Sylvian fissure, obliterated sulci or widened sulci, infarction, temporal horn enlargement, periventricular hypo-density, unequal temporal lobe or unequal temporal fossa, excessively thickened skull, abnormal shape of the skull, very small and thick temporal bone, occipital horn enlargement, unilateral ventriculomegaly, smaller hemicranium and normal pressure hydrocephallous.[6] A compact brain means the obliteration of all sulci except widened Sylvain fissure. In CT scan we can find very small ventricle, alternately enlargement of ventricle due to parenchymal tissue loss, brain atrophy, hypo-density in cingulated gyrus. A postoperative CT scan is also required to get an idea of the post-operative improvement and guidance. Growth, nutrition and physical activity are important determinants of health outcomes in children with CP.[7] So balanced diet with high protein diet also essential part of CP treatment along with cranioplasty and duraplasty. Management of CP requires the involvement of multispecialty or multidisciplinary team members. Most of the patients suffering from CP are from poor families. Parents of those children are not even able to provide nutritious food. Hence, it is impossible for them to treat their children with such a physician team. Those who have financial ability can take multidisciplinary management support. Usually, the neuro-physician or paediatrics consultant who used to treat CP patients is the team leader who makes a plan for long-term treatments. Orthopedists and spine surgeons look for preventing contractures, hip dislocations, and scoliosis. Physical therapists develop and implement care plans to improve movement and strength, and administer formal gait analyses. Occupational therapists develop and implement care plans focused on activities of daily living. Speech and language therapists develop and implement care plans to optimize the patient's capacity for communication. Surgical procedures may improve mobility and decrease pain. Surgical management options include placement of a baclofen pump, selective dorsal rhizotomy, tendon releases, hip dislocation/rotation surgery, spinal fusion, strabismus repair, and deep brain stimulation.[8-9]

In this case, cranial surgery was done. The Aim of this surgery was to increase the volume of the cranial cavity. To achieve this goal bilateral cranioplasty and duraplasty were done. During surgery, it was found that the cranial bones of this patient were thicker than normal or even thicker than the bones in adults. Temporal bones were thickened and the dural pulsation was absent. After duraplasty, dural pulsation appeared. In the postoperative period, the patient usually requires intense monitoring, pain relief, and maintenance of adequate intravascular fluid volume and electrolytes level. However, in this case, the patient recovered from anesthesia smoothly. Just after recovery from anesthesia, in the postoperative ward, the movement of her limbs were forceful and able to grip. Her spasticity subsided,and her convulsion disappeared as well. After two hours, an increased feeding tendency was observed. Her response to her mother's call was noticed too. Her weight also gradually increasing. A CT scan of the brain was done on 2nd postoperative day. Though the brain looks very healthy, a lot of collection of CSF was observed that was compressing the brain from all sides of the brain. On the 5th postoperative day,CSFdrainage was done. But 3 to 4 days following the drainage, again CSF collection took place again. A CT scan of the Brain was done again and it was the same as the previous

CT scan report. Then a search for the dural gap was done following the opening the wound under general anesthesia. Finally, the gap was identified and repaired. For the first week of 2nd time post-operative care, the patient was losing weight. After 4 to 5 days apatite of the patient was regained. The patient is now completely normal and rapidly growing with a smiling face. Hence,cranial surgery could be considered the real solution of curable CP treatment.

Prognosis with usual treatment

Usually, CP patients survive up to adulthood. Most common causes of the early death of CP patients are respiratory diseases like aspiration pneumonia, choking, deglutition failure, and repeated respiratory tract infection due to poor dental hygiene and poor nutrition[10] CP patients grow up to adults. About 90% of CP children survive until 20 years. While 95% of CP patients with diplegia and 75% of CP patients with quadriplegia survive up to 30 years. It has also been reported that 95%of CP patients who have mild cognitive deficits and 65% of CP patients with severe cognitive deficits survive until the age of 38 years.[11] If the CP patients have seizures, results will be worst. Actually, no curative treatment is available to treat CP and multidisciplinary approach treatment makes it very expensive. Because of the economic condition of the parents, most of the CP patients are being treated with physiotherapy, speech therapy, and tendon transfer. CP patients are economic, social, and psychological burdens to the whole family. In addition, if CP patients have bowel bladder incontinence, the life of them and their parents become miserable. Hence, early diagnosis and early cranial surgery could save CP patients life and their families.

Prognosis with invented cranial surgery

Immediate changes observed in early postoperative period with the forceful movement of her all limbs. She was trying to remove the I/V cannula, which was placed in her distal forearm. That means now she has gained the capacity to grip strongly. The patient's mother excitedly and emotionally was demanding to take the baby in her lap. Her mother was allowed to do that and after taking her on her lap the patient gripped her mother's cloth. The mother was calling the baby and she was responding. One month later the patient came for a follow-up visit and the patient holding up a colourful plastic flower which indicated that her vision was also improving.

Conclusion

From the outcomes of this case study, it could be concluded that bilateral cranioplasty and duraplasty (Tanfarid Procedure)[12] could be the curative surgical treatment for CP as the patient responded positively just after recovery from anesthesia. Others available treatment options are not be able to cure CP. Rather if conventional treatment will continue in this patient there is the possibility of the development of permanent blindness, deformity of limbs and ultimately early deglutition failure and respiratory failure. Now the patient is able to see and pick up toys herself as she likes.

Acknowledgment

The authors are thankful to the "Neurodevelopmental disorder welfare association" for their support.

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