

Physiotherapeutic Management of Hydrocephalus-Ex Vacuo: A Case Report

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ABSTRACT

Hydrocephalus Ex-vacuo refers to an elevated volume with no increasing Cerebrospinal Fluid (CSF) pressure, particularly in instances of diminished brain tissue. Since there is no increased intracranial pressure in Hydrocephalus Ex-vacuo, few general symptoms such as irritability, drowsiness, and even larger head presentation can be absent compared to congenital hydrocephalus. The current case study presents a one-year-old male child exhibiting delays in developmental milestones, including poor head and trunk control, associated with a history of prenatal bleeding and subsequent birth complications. Despite a seemingly normal birth, the child experienced postnatal complications such as vomiting, seizures, and abdominal distension, leading to a 15-day Neonatal Intensive Care Unit (NICU) stay. Persistent delays prompted medical consultations, culminating in a referral for physiotherapy following Magnetic Resonance Imaging (MRI) findings indicative of hydrocephalus ex-vacuo. Clinical assessments revealed a restricted range of motion, muscle tightness in the lower extremities, and poor muscle control and reflex persistence. Imaging confirmed ventricular enlargement consistent with Hydrocephalus Ex-vacuo, necessitating targeted physiotherapy interventions. Therapeutic strategies focused on promoting head and trunk control through prone positioning, bolster support, and dynamic exercises utilising physio balls. Progressive interventions facilitated significant improvements within six months, highlighting the efficacy of early rehabilitation and parental involvement in achieving motor milestones. The present case underscores the importance of timely diagnosis, multidisciplinary collaboration, and rehabilitative interventions in optimising outcomes for children with hydrocephalus ex-vacuo. Further research is warranted to explore additional therapeutic modalities and long-term prognostic indicators for this patient population.

Keywords: Congenital, Developmental delay, Infant, Physical therapy, Ventricular enlargement

CASE REPORT

A one-year-old male child was brought to the Outpatient Department (OPD) by his mother, who complained that her child was unable to lift his head, turn around on his own, sit, stand, and make eye contact like other children of his age. The mother had a history of bleeding in the 7th month of pregnancy and was admitted to the hospital for one day. The mother experienced pain on 21st September 2020, i.e., 37 weeks and three days of gestation. The baby boy was delivered by vacuum-assisted normal delivery on 22nd September 2020 at 1:45 pm with a birth weight of 3kg, a good cry, and Appearance, Pulse, Grimace, Activity and Respiration (APGAR) scores of 7/10 at one minute, 9/10 at five minutes. Immediately on the next day, the child had a history of vomiting and abdominal distension, with one episode of seizure. The baby was under observation in the NICU for 15 days. Gavage feeding was done for three days, and later paladai feeding was started. Breastfeeding was initiated on the 10th day. Delays in milestones were noted by the mother around 2-3 months. Hence, she consulted at multiple medical facilities, and no changes were noticed in the child's status. The child was referred to physiotherapy after an MRI during the ninth month of life revealed ex-vacuo dilatation of the posterior horn of the lateral ventricles. A higher mental function examination suggests that the youngster was awake and alert. The kid attempts to socialise by cooing and making specific noises. These language abilities were attained at the age of 3-4 months. Furthermore, the child approaches the mother and relatives with a smile, and weeps or shows fright when entering the OPD or seeing the physical therapist, demonstrating social communication skills and memory power. This indicates the child's ability to remember and recognise known people. The auditory and visual functions appear normal and were confirmed by the head turning to auditory and visual stimuli such as light and colourful toys.

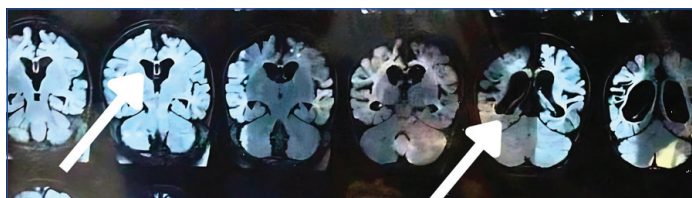
On observation in the supine position, the hips are slightly flexed and externally rotated, knees flexed, and ankles plantarflexed. The head is rotated to the right-side, the shoulders are slightly abducted, fingers are flexed bilaterally. The child was not initiating rolling to any side. When the baby was placed in a prone position, he could not tolerate the position for more than a minute. No head lifting or initiation to roll back to supine was noted. Fine motor and gross motor milestones were not achieved. The persistence of all spinal primitive reflexes was noted, such as flexor withdrawal, crossed extension, extension thrust, Moro's reflex, and palmar and plantar reflexes. Rooting and sucking reflexes were integrated. Brainstem reflexes like asymmetrical tonic neck reflex, symmetrical tonic neck reflex, and tonic labyrinthine reflex were persisting.

The baby reacted to touch, pain, sound, and light, indicative of a well-functioning sensory system. Passive Range of Motion (ROM) was measured for all joints using a Goniometer, revealing restricted ROM in the bilateral lower extremities. Muscle tightness was noted in the hamstring and calf muscles. Based on Daniels and Worthingham's muscle testing, the baby exhibited fair power in the bilateral upper and lower limbs. The movements performed by the child were mostly parallel to the ground, indicating weakness and poor muscle control against gravity. Muscle tone was assessed using the Modified Ashworth Scale (MAS) [1,2]. A slight increase in muscle tone was noted in all four limbs, i.e., grade 1 for elbow flexors, hip flexors, and knee flexors bilaterally.

Few special tests were performed on the child. Firstly, head lag was noticed during the pull-to-sit test [3], indicating poor head and neck control. In the Hamstring – 90° – 90° method, knee extension test [4,5], a limited range was noted. A 30-degree angle on the left-side and a 35-degree angle on the right-side were observed.

Lastly, the Calf-Silfverskoild test [6,7] was performed. It was noted that dorsiflexion without knee flexion was more restricted than dorsiflexion with knee flexion.

Echocardiography showed a small 5 mm Atrial Septal Defect (ASD) with a left-to-right shunt. Furthermore, Brain MRI (plain) revealed bilateral parieto-occipital and perirolandic T2 hyperintensities partially suppressing on Fluid-attenuated Inversion Recovery (FLAIR), with parenchymal volume loss and hydrocephalus ex-vacuo dilatation of the posterior horn of lateral ventricles. No evidence of diffusion restriction or haemorrhagic foci was found. The rest of the cerebral parenchyma showed normal signal appearance (myelination pattern corresponding to the age of the subject). Basal ganglia, thalami, internal capsules, and Corpus callosum appeared normal. The rest of the lateral ventricles and the third ventricle appeared normal. Additionally, benign prominence of cortical sulci was noted. Sella, parasellar cavernous sinus, and suprasellar regions were normal. No mass lesion or midline shift was evident in the scan. In conclusion, the major MRI findings were bilateral parieto-occipital and perirolandic T2 hyperintensities partially suppressed on FLAIR, with parenchymal volume loss and Hydrocephalus Ex-vacuo dilatation of the posterior horn of the lateral ventricles, cystic encephalomalacic changes (sequelae of neonatal Hypoxic Ischaemic Encephalopathy (HIE)) [Table/Fig-1].











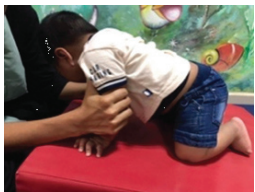
[Table/Fig-1]: Brain MRI (dilatation of posterior horn of lateral ventricles).

The International Classification of Functioning, Disability, and Health (ICF) Model was used to classify body structure and function abnormalities such as reduced ROM in bilateral legs, poor head and neck control, and trunk control. Muscular power decreased, but muscular tone increased. In terms of participation restrictions, the baby had trouble engaging in regular age-appropriate exploration of the environment.

The physiotherapy was designed to achieve developmental milestones such as turning over, sitting, and crawling while also aiming to improve control of the head and neck. Additionally, visual training for eye tracking and fixation was provided.

The physiotherapy session commenced when the child was one year and 10 days old and referred to the Paediatric Physical Therapy Department. Details are provided in [Table/Fig-2(a-i)].

Aim	Treatment strategies
To improve head and neck control  <p>a) Bouncing on the physio ball by stabilising the head and neck.</p>	Bouncing on the physio ball by stabilising the head and neck The therapist's hand stabilises the child's head from the Vertex/top of the head. Mild downward compression was given in between, a frequency of 2-3 times with the neck in the neutral position, to activate cervical joint proprioception for facilitating muscle activities.
 <p>b) Prone on peanut ball.</p>	Home exercises: The mother was advised to stroke on the posterior aspect of the neck and to perform the above exercises at home. <ul style="list-style-type: none"> Prone on peanut ball-rolling forward-wait for the child to lift the head against gravity. Duration: <ul style="list-style-type: none"> Total 10 minutes of training for head and neck 1-2 minutes of activity followed by rest for 2 minutes.

To improve trunk mobility  <p>c) Trunk rotation in supine position.</p>	Stretching and trunk rotation in sitting and supine Trunk rotation in a sitting position. The child's pelvis was stabilised and then the upper body turned to the right and left direction with a 30 sec to one minutes hold at the end range. Supine stabilising pelvic and rotating the trunk to both direction and vice versa (Stabilising the trunk and rotating hip to both side) Each exercise performed three sets to both side.
To facilitate rolling  <p>d) Rolling over physio ball. (Images from left to right)</p>	Rolling over physio ball. (Dynamic surface) <ol style="list-style-type: none"> Stabilising the lower body Stabilising the upper body The exercise was performed on the mat initially (Static surface) 1-2 minutes of activity followed by rest for two mint. Each side turning trained for 5-8 minutes.
To improve trunk control  <p>e) Bouncing over the physio ball. (Images from left to right)</p>	<ol style="list-style-type: none"> Bouncing over the Physio ball with minimal support over the waist. (Bouncing to activate more joint proprioception as well as a playful activity to keep the babies attention on sitting) Core activation exercise on physio ball by moving the ball in all directions. E.g., Moving the baby backward on the ball and wait for the child reaction to come forward by activating the trunk/core muscles. 1-2 minutes of activity followed by rest for two minutes. Each side turning trained for 5-8 minutes.
To facilitate quadruped position  <p>f) a) weight bearing on upper limb; b) Prone on elbow. (Images from left to right)</p>	To promote elbow extension and to improve upper limb strength <ol style="list-style-type: none"> Prone on elbow, progressing to prone on hand and Sitting weight bearing on upper limb using wedge and bolster 2-3 minutes of sustained posture followed by 1-2 minutes rest Total duration of exercise 15-20 minutes.
To promote hip control and improving gluteal strength  <p>g) Iliopsoas activation.</p>	<ol style="list-style-type: none"> Iliopsoas activation- Upper body supported on bolster and child's knee and thigh supported by therapist lap. The trunk was suspended. Therapist hand under the tummy to support drop. Iliopsoas will be stretched whenever child drops the hip down. To activate the muscles mild abdominal squeeze was given. Which facilitate child to lift the trunk up. Kneel sitting to kneel standing-hands supported on bolster. Quadruped position with support 2-3 minutes of activity followed by 1-2 minutes rest Total duration of exercise 15-20 minutes.
 <p>h) Kneel sitting.</p>	
 <p>i) Quadruped position.</p>	

To improve tracking of objects	As child enjoyed bright lights, child was placed in a room with no light, and visual tracking was stimulated by displaying flash light on the ceiling/ Wall of the room. Different plastic colour filter to produce different colours of light. Home exercises-The mother was suggested to perform the same technique at home.
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[Table/Fig-2a-i]: Intervention details.

After six months of physiotherapy, the child had regained head and neck control and the ability to roll. The child could sit comfortably with support, and tracking abilities improved due to faster eye and neck movements.

DISCUSSION

Hydrocephalus ex-vacuo is a condition characterised by increased CSF within the cranial cavity, often as a compensatory mechanism due to brain atrophy or loss of brain tissue. The present case report presents a one-year-old child exhibiting poor head and trunk control, common symptoms associated with hydrocephalus ex-vacuo. Early recognition and comprehensive rehabilitation can significantly impact the quality of life for these patients. According to numerous studies, the prevalence of hydrocephalus in the paediatric population ranges from 30 to 423 per 100,000. Congenital hydrocephalus occurs in approximately 0.2 to 0.5 out of every 1,000 live births. However, the incidence of hydrocephalus ex-vacuo is seldom reported due to its infrequent occurrence [8,9].

The prenatal and perinatal factors play a crucial role in understanding the underlying aetiology of the child's condition. Maternal bleeding during the seventh month of pregnancy might have led to foetal distress, potentially affecting the baby's brain development and resulting in hydrocephalus [10,11]. The vacuum-assisted normal delivery could also contribute, albeit not directly causative [12,13].

The clinical presentation of poor head and trunk control in infants often raises concerns regarding neurological dysfunction. Imaging studies, such as MRI or Computed Tomography (CT) scans, play a crucial role in confirming the diagnosis of hydrocephalus ex-vacuo [14,15]. In the present case, imaging revealed ventricular enlargement consistent with the condition, indicating compensatory CSF accumulation secondary to brain parenchymal loss.

Head control is the first developmental milestone achieved by humans. If not achieved, it can delay the remaining milestones. Therefore, prioritise achieving head control initially. Prone positions like tummy time were used to improve head control [3,16]. The baby was trained over a bolster, supported at the trunk, and over the physio ball to train static and dynamic activity. The activity progressed to quadruped, where baby head control was facilitated without tummy/trunk support. Studies have reported that the Swiss ball exercise is found to be useful in training and could improve trunk control, balance, and motor skills in spastic children. Similar techniques to improve trunk muscle control and balance [17-20].

The prognosis for children with hydrocephalus largely depends on the extent of brain damage, the effectiveness of treatment, and the availability of supportive interventions [12,21]. Long-term follow-up is essential to monitor developmental progress, address emerging challenges, and modify therapeutic strategies accordingly [22].

The parents were advised to continue the home program and exercise, which helped us achieve the outcome as soon as possible. In the present study, the child attained head and trunk control within six months. This study promises us that early rehabilitation [23,24] and parent participation will help children with hydrocephalus ex-vacuo to achieve their motor milestones.

CONCLUSION(S)

The present case report illustrates the clinical presentation, diagnosis, and management of hydrocephalus ex-vacuo in a one-year-old child presenting with poor head and trunk control. Through timely diagnosis and appropriate intervention, including physiotherapy, significant improvements in motor function were achieved, leading to enhanced head and trunk control and the ability to sit independently. The present case underscores the importance of early recognition, multidisciplinary collaboration, and rehabilitation in optimising outcomes for children with hydrocephalus ex-vacuo. Further research and clinical studies are warranted to explore additional therapeutic interventions and long-term outcomes in the present patient population.

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