



Case Study of a Child with Sturge Weber Syndrome

Begum R*

Senior Clinical Physiotherapist & Incharge of Paediatric Physiotherapy Services, Paediatrics Department, Centre for the Rehabilitation of the Paralysed (CRP), Savar, Dhaka, Bangladesh

*Corresponding author: Begum R, Senior Clinical Physiotherapist & Incharge of Paediatric Physiotherapy Services, Paediatrics Department, Centre for the Rehabilitation of the Paralysed (CRP), Savar, Dhaka, Bangladesh; Tel: + 8801675793257; E-mail: physio_miilee@yahoo.com, [drabea3\[at\]gmail\[dot\]com](mailto:drabea3@gmail.com)

Abstract

Sturge-Weber Syndrome is a rare, progressive and congenital syndrome. This syndrome is characterized by a facial birthmark and neurological abnormalities with port wine stain, developmental delays, glaucoma, seizures and paralysis. The aim of this case is to present the developmental delay of Sturge-Weber syndrome (SWS) with physiotherapy management. A 9 month old girl was diagnosed as SWS. The boy was examined by MRI scan. This patient was admitted at Paediatric Department for 14 days. Physiotherapist focused child's developmental milestone and prevention of complication. Early identification and physiotherapy treatment plan is important to prevent development of further complications.

Keywords: Sturge-Weber syndrome; Physiotherapy; Milestone

Introduction

Sturge-Weber Syndrome is rare congenital disorder with vascular malformations in the skin, eye and the brain. Thus these malformations result from failure of normal regression of fetal vascular plexus surrounding the cephalic portion of the neural tube [1]. The ectoderm over this area later forms the facial skin. The prevalence is actually estimated at 1 per 50,000 live births [2]. Sturge Weber Syndrome (SWS) was first described by Schirmer in 1860 and later more comprehensively by Sturge in 1879 who associated dermatological and ophthalmological changes of the disease to the neurological manifestations. Weber complemented it with the documentation of radiologic findings seen in these patients [3]. The aim of the study is to show the developmental delay of child with SWS with physiotherapy management to improve developmental milestone age appropriately.

Case Description

Subjective information

A female infant came to inpatient, Paediatric department at CRP, Savar on 15th July, 2020. She was 9 months old and her ID no. was 20/15827. She was from Faridpur district of Bangladesh. The referring diagnosis was "Sturge weber syndrome". Her parents are well educated, both of them completed post-graduation. Her mother is a housewife and father is a teacher. She has no siblings. Her mother came with the complaint that she couldn't hold her neck and sit independently. Her parents' expectation was to recover her condition as much as possible. After explaining about the condition, she had given a written consent to complete the child's entire assessment and therapeutic intervention here at inpatient pediatric department, CRP, Savar, Dhaka.

From the birth history author found that she was a term child, the delivery of the child was attended by a doctor at hospital. After birth she had pneumonia, and a seizure was seen after eight hours of birth. They stayed for four days at the hospital and received conservative treatment (oxygen and medication). The investigation report of EEG and CT-scan were available. The report of EEG was abnormal due to the presence of frequent slow waves in the right temporal region which is consistent with focal

Received date: 26 August 2020; **Accepted date:** 02 September 2020; **Published date:** 07 September 2020

Citation: Begum R. (2020). Case Study of a Child with Sturge Weber Syndrome. SunText Rev Case Rep Image 1(1): 104.

DOI: <https://doi.org/10.51737/2766-4589.2020.004>

Copyright: © 2020 Begum R. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.



cerebral dysfunction. And the impression of the CT-scan showed that she had Sturge Weber Syndrome. She had taken homeopathy treatment before and had been continuing medication for epilepsy. Her last seizure was seen at 7.5 month of her age. She had no hearing problem but had a fixing vision and communicated with her mother by crying. For ensuring the accessibility, I had to know about the child's home environment and the paved road was pitched and type of area was urban.

Objective information

By general observation I found that the child was lying on her mother's lap. I observed that the respiratory status of the child was normal (respiratory rate). A mild increased tone was present on both of her upper and lower limbs. The ROM of both of her upper and lower limb was in within normal limit. The ROM of spine was also within normal limit. She had symmetrical but poor weight bearing in sitting position. And she had poor protective and equilibrium reaction in sitting position. Poor balance & co-ordination were also present.

In gross motor function, I found that the main mode of mobility for the child was lying (dependent on her mother) and the preferred position of floor for the child was supine lying. The child needed total support on bridging, cross leg sitting, long sitting, box sitting, four-point kneeling, squatting, high kneeling and standing. In case of transitional movement, she could perform roll over from prone lying to supine lying position. This Maximum support was needed for lying to sit. Her fine motor function and coordination of upper limb was poor.

After completing subjective and objective assessment physiotherapist diagnosed this patient as Sturge Weber Syndrome.

Treatment plan and goal set-up

After completing subjective and objective assessment, some problems were found out to be treated. Developmental day was focused point to discuss. The physiotherapy problems included: increased muscle tone, poor posture, poor gross motor function, poor transitional movement (lying to sitting, sitting to standing), poor coordination, poor grasping and releasing object. According to problem physiotherapist fixed physiotherapy treatment plan with short term and long term goal. The treatment plan was following:

- facilitate active range of motion exercise to improve active movement
- passive range of motion exercise to prevent contracture and deformity
- sustained stretching to reduce muscle tone in upper and lower limb

- functional stretching to improve movement, flexibility through neuroplasticity
- pelvic control practice by bridging exercise
- practice gross motor function to improve or achieve milestones
- facilitate transitional movement to improve mobility
- postural correction
- mother's counseling about child's condition, prognosis, future complications and importance of physiotherapy

Therefore, I had arranged some short term and long term goals according to the child's problem list, and set duration to achieve those goals. Those short term goals were to provide mother's education, reduce tonicity, improve neck control and improve transitional movement (supine to prone) within 14 days. And long term goals were to improve neck control in sitting, improve posture, improve mobility through within 4 months and advised child's mother to come over at CRP for follow-up of her child from time to time.

To achieve these goals, I had to come up with some treatment plans. I planned to provide slow passive stretching, active facilitating exercise and weight bearing to reduce tonicity. I had also planned to provide neck control practice in sitting and prone lying to improve neck control, bridging exercise to improve pelvic control, rolling practice to improve transitional movement, gross motor function practice (according to severity) to improve gross motor function, postural correction to improve posture, sitting balance practice to improve sitting balance, and mother's education about patient's condition, treatment, prognosis.

I also maintained SOAP note regularly and had given home advice to the mother of the child to prevent secondary complications. The child was discharged after only one week, on 23.07.200 that is before the expected time due to several attack of convulsions. She was discharged and referred to Dhaka Medical College (DMC) for management of convulsion. By the time of discharge, the mother was completely educated about her child's condition, treatment, handling, prognosis and the importance of physiotherapy for her child. But her condition was not so much improved. Child's mother was advised to perform all group therapies, hydrotherapy, physiotherapy accurately and regularly by maintaining dose and instructed not to perform therapy if the child had fever or got sick, rather consult with a doctor for betterment of the child's conditions. Wooden chair was planned to prescribe for improving static sitting balance and to maintain body alignment for postural correction.

Discussion

A study with Sturge Weber Syndrome exhibited the prevalence of clinical manifestation and relationship among variables. The age of onset of seizures was (0-23 years) and the relationship between



the seizures and developmental delay was established. In those with and without seizures, the prevalence of developmental delay was (43% vs. 0%), emotional and behavior problems (85% vs. 58%), special education requirements (71% vs. 0%), and employability (46% vs. 78%) were analyzed [4]. The resent study showed similar feature of developmental delay with feature. Due to methodological design author was not able to analyze the relationship between seizure and developmental delay. Another case with SWS developed seizure at age of 2 year and seizure respond well to the antiepileptic treatment. This patient did not have development delays and higher functions were normal [5].

Conclusion

Sturge-Weber syndrome illustrates a various types of clinical presentations where seizure and developmental delay are common features. Physiotherapy management is necessary to avoid future complications and to improve developmental milestones of patients.

References

1. Fatai MA, Mary AO, Adenike AO. Sturge-weber syndrome: a case report in a 39 yr- old man with delayed diagnosis. *Austin J Clin Neurol*. 2015; 2: 1049.
2. Di Rocco C, Tamburrini G. Sturge–Weber syndrome. *Child's nervous system*. 2006; 22: 909.
3. Neto FXP, Junior MAV, Ximanes LS, de Souza Jacob CC, Junior AGR, Paletha ACP. Clinical features of Sturge Weber Syndrome. *Intl Arch Otorhinolaryngol*. 2008; 12: 565-570.
4. Sujansky E, Conradi S. Outcome of Sturge-Weber syndrome in 52 adults. *Am J Med Genet*; 1995; 57: 35-45.
5. Nigam P, Thakare S, Singh UP, Indurkar M. Sturge-Weber Syndrome: A Case Report. *Sch J App Med Sci*. 2017; 5: 3060-3064.