



Case report

Rehabilitation of stroke in cyanotic congenital single ventricle heart defect patient

Ragini Dadgal, Mohd. Irshad Qureshi, Divya Jethwani, Waqar M. Naqvi*, Chaitanya A. Kulkarni

Ravi Nair Physiotherapy College, Datta Meghe Institute of Medical Sciences, Wardha, Maharashtra, India

ABSTRACT

Congenital heart disease consists of various conditions including tetralogy of Fallot, ventricular septal defect, Epstein's anomaly, single ventricle, etc. Among these single ventricles is one of the gravest forms of cyanotic congenital heart disease. The cardiac diagnosis is associated with an increased risk of stroke among children. Pediatric arterial ischemic stroke (AIS) is an important cause of neurologic disease in children causing disability. The 14-year-old patient came to the hospital was presented with left side hemiplegia with severe exercise intolerance due to congenital heart disease. The patient has been advised to undergo Fontan procedure for single-ventricle condition 3 years back, but due to poor socioeconomic status, parents of patients refused to do so. The primary goal was to improve bed mobility and trunk balance without developing symptoms of exercise intolerance. The intervention was started with deep and segmental breathing exercises. Proprioceptive neuromuscular facilitation and constrained induced movement therapy were added in the program in addition to passive and active movements, bed mobility, functional reeducation, trunk control exercises, and balance exercises. Combinations of all of the above therapeutic approaches lead to increased functional independence in the patient. This case reports the effectiveness of a rehabilitation program for pediatric arterial ischemic stroke with preventive guidelines for exercise intolerance.

Keywords: Congenital heart disease, exercise intolerance, pediatric stroke, Physiotherapy interventions

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Correspondence: Waqar M. Naqvi* ✉ waqar.naqvi@dmimsu.edu.in

Department of Community Health Physiotherapy, Ravi Nair Physiotherapy College, Datta Meghe Institute of Medical Sciences, Wardha, Maharashtra, India.

INTRODUCTION

Congenital heart disease consists of various conditions including tetralogy of Fallot, ventricular septal defect, Epstein's anomaly, single ventricle, etc. Cardiac diagnosis is associated with an increased risk of stroke [1]. Among these, Single ventricle is one of the gravest forms of cyanotic congenital heart disease [2]. "A single ventricle is defined by a heart with inflow from two separate atrioventricular valves or a common atrioventricular valve emptying into one ventricle". It has been shown that the most common cyanotic CHD are due to the transposition of the great arteries [3].

There is a high prevalence of arterial ischemic stroke in children with cardiac disease. Pediatric arterial ischemic stroke (AIS) is an important cause of neurologic disease in children causing disability. Consequences can include sensorimotor deficits, language impairment, intellectual disability, behavioral problems, and epilepsy. Although many cases of AIS have a multifactorial etiology, cardiac disorders are associated with 10% to 30% of strokes in children [4].

The purpose of the case report was to describe the case of cyanotic congenital heart disease associated with pediatric stroke, to

plan the interventional strategy, and to describe the outcomes.

Case presentation

A 14-year-old girl came to the OPD with a chief complaint of left side weakness. She is a known case of congenital heart disease and underwent Glenn shunt for single ventricle condition in 2011, was brought with complaint of weakness on the left side of the body since 10th May 2020. She had peripheral cyanosis, clubbing (grade 3). On auscultation, she had a systolic murmur. Left-sided hypotonia was present.

MRI brain was done which was s/o acute infarcts in right high parietal and frontal regions and chronic infarcts in right ganglio-capsular and corona radiata regions. 2D Echo was done which was sign of Double inlet left ventricle with malposed greater arteries, normal ventricular function.

Patient's lives in a small house. Does not go to school due to exercise intolerance, is independent in ADLs. With the chief complaint of the patient's parents, she is unable to sit, feels lethargic with dizziness on changing positions in bed.

On Evaluation

Table 1: Deep Tendon Reflexes

| Right | Reflex | Left (1 st Week) |
|--------|-----------|-----------------------------|
| Normal | Biceps | 2+ |
| Normal | Triceps | 2+ |
| Normal | Supinator | 2+ |
| Normal | Knee | 2+ |
| Normal | Ankle | 2+ |

Table 2: Spasticity as per Modified Ashworth scale

| Muscles (left) | 1 st week | 2-3 weeks | 4-5 weeks |
|---------------------------|----------------------|-----------|-----------|
| Shoulder flexors | 0 | 1+ | Normal |
| Shoulder abductors | 0 | 1+ | 1 |
| Shoulder medial rotators | 0 | 1+ | 1 |
| Shoulder lateral rotators | 0 | 0 | Normal |
| Elbow flexors | 0 | 1+ | 1 |
| Wrist flexors | 0 | 1 | 1 |
| Hip flexors | 0 | 1+ | Normal |
| Hip lateral rotators | 0 | 1+ | 1 |
| Knee flexors | 0 | 1+ | 1 |
| Ankle Dorsiflexion | 0 | 1 | Normal |

Table 3: Voluntary motor control

| Group of muscles | 1 st week | 2-3 weeks | 4-5 weeks |
|------------------|----------------------|-----------|-----------|
| Shoulder | 2 | 3 | 4 |
| Elbow | 2 | 3 | 5 |
| Wrist | 1 | 2 | 4 |
| Hip | 1 | 2 | 4 |
| Knee | 1 | 2 | 4 |
| Ankle | 1 | 2 | 4 |

Joint Integrity

Passive ROM is full in the bilateral upper extremity and lower extremity.

Table 4: Co-ordination Tests

| Left | Non-equilibrium | Right |
|------|----------------------|-------|
| Poor | Finger to nose | Good |
| Poor | Finger to finger | Good |
| Poor | Heel to shin | Good |
| Poor | Pronation supination | Good |

Table 5: Hand Functions

| | |
|---|----------------|
| Grasp a) spherical b) cylindrical c) hook | No No No |
| Grip a) pulp to pulp b) tip to tip | No No |

| | |
|---|----------|
| c) three jaw chuck d) lateral prehension | No No |
|---|----------|

Diagnostic Scales

To measure the outcome after treatment, 2 scales were used.

1. Pediatric Balance Scale: The pretreatment score was 0 out of 56.
2. Postural assessment scale for stroke: Pretreatment score was 5 out of a maximum score of 36.
3. FIM: The pretreatment score was 40 out of 126.

Diagnostic challenges

The patient has been advised to undergo Fontan procedure for single-ventricle condition 3 years back, but due to poor socioeconomic status, parents of patients refused to do so. So, the underlying heart condition remains untreated. In addition, due to lack of public transport and risk of COVID-19 infection, the patient's parents refused to participate in further rehabilitation on an OPD basis. Exercise intolerance also reduces the patient's motivation to participate in exercise programs.

Week 1: Early physiotherapeutic intervention was started 2 days after the patient was admitted to the ICU of the hospital. As the patient is suffering from cyanotic congenital heart disease, the neuro-physiotherapy intervention was started keeping in mind exercise intolerance of the patient. Reflexes were found to be exaggerated on evaluation (Table 1). Cardiovascular exercise testing could not be done. The primary goal was to improve bed mobility and trunk balance without developing symptoms of exercise intolerance. The intervention was started with deep and segmental breathing exercises. Passive arm and leg exercises accompanied by deep breathing were included in the program to increase oxygen uptake and reduce exercise intolerance [5]. Stretching exercises were integrated to prevent restricted joint mobility [1]. Precautions were taken such that patients do not have to perform any static exercise. The patient was instructed to avoid the Valsalva maneuver during any exercise. 5 repetitions interspersed with 20 second rest period with pursed-lip breathing exercises were started and increased over the period of time as the patient's capacity increased.

Ankle pumping exercise was advised to avoid thrombosis. Active exercises were started for the right upper and lower extremities. Left hip flexion-extension, adduction abduction, knee flexion-extension movements were given supported by the therapist in gravity eliminated plane and active assisted exercises for left upper extremity to develop motor control. Bed mobility exercises like rolling, bridging, supine to sitting were started with moderate support. Electric muscle stimulation was given to Deltoid, scapular muscles, triceps to avoid painful shoulder and to quadriceps, ankle dorsiflexion, plantar flexors to stimulate muscle activity [2].

Week 2-3

Active assisted exercises were started for left lower extremity, left wrist, and hand which was progressed to active

exercises and resisted exercises for the upper extremity. Sitting balance exercises and trunk control exercises were also started. All exercises were interspersed with 20 second rest period and pursed-lip breathing exercise. Supported standing exercises were started with the hand reaching out to the tabletop on the right side. The patient could stand for 10 seconds after which she started feeling dizzy with an increase in heart rate. Therefore standing for 5 seconds with 20 seconds rest period for repetition up to 5 times was started. Constrained induced movement therapy was included in the program for 30 mins a day for improving hand function (Table 5).

Week 4-5

The patient is now stable and shifted to a general pediatric ward from ICU. Active exercises with minimal manual resistance for bilateral upper and lower extremity could be done. The patient could stand independently and the period of standing could be increased to 15 seconds with an interspersed rest period. Heel raising in standing and step up and step down was started for 15 seconds. All other exercises were continued. As the patient's capacity was found to be increased by a small amount Proprioceptive neuromuscular facilitation was started.

Motor control was improved significantly after 5 weeks of physiotherapy intervention and the patient became independent in activities of daily living. She could walk in the room and climb stairs with moderate support. After 5 weeks pre and post-assessment were compared. The tone changed from grade 0 i.e. flaccidity in the first day to grade 1 to normal up to end of the 5th week (Table 2). Voluntary Motor control changed from grade 2 to grade 4 (Table 3). Coordination and balance improved from poor to fair (Table 4). The pediatric balance scale score was improved from 0 to 46 out of 50. The postural assessment scale for stroke (PASS) score was improved from 0 to 34 out of 36. FIM was improved from 40 to 97 out of 126.

DISCUSSION

The patient's congenital cardiac anomaly of Double inlet left ventricle with malposed greater arteries leads to cyanosis and pulmonary hypertension. Patients with this condition are prone to exercise intolerance^[5].

The early physiotherapeutic intervention was started 2 days after the patient was admitted in the ICU of the hospital. It is mentioned in many studies that the early start of rehabilitation causes better recovery and leads to better results^[6]. Pursed lip breathing, deep and segmental breathing exercises, passive arm and leg exercise accompanied by deep breathing were included in the program to increase oxygen uptake and reduce exercise intolerance^[7]. Precautions were taken such that patients do not have to perform any static exercise^[8]. The patient was instructed to avoid Valsalva maneuver during any exercise or during any daily activity as it is proven to be causing an increase in the intrathoracic pressure^[9]. All

these techniques and precautions were found to be helpful as it helped in preventing dyspnea and in turn preventing the anxiety in the patient-related to physiotherapy session. This leads to better patient cooperation and better recovery. Proprioceptive neuromuscular facilitation and constrained induced movement therapy were added in the program in addition to passive and active movements, bed mobility, functional reeducation, trunk control exercises, and balance exercises. Combinations of all of the above therapeutic approaches lead to increased functional independence in the patient^[3-6].

CONCLUSION

This case report includes a rehabilitation program for cyanotic congenital single ventricle heart defect patient having pediatric arterial ischemic stroke with preventive guidelines for exercise intolerance.

LIMITATION

As the patient is known case of cyanotic congenital heart disease, she was not able to reach her full potential.

AUTHORS' CONTRIBUTIONS

RD undertook the case and reported it. WMN and CAK assisted in manuscript preparation. WMN, IQ, DJ approved the manuscript for publication.

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DECLARATION OF PATIENT CONSENT

The authors certify that appropriate consent forms were obtained from the patient for preparing the case report.

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