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# Inpatient cardiac and respiratory physiotherapy management of a child with Down syndrome who underwent surgical repair of congenital heart disease: A case report

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## ABSTRACT

Down syndrome (DS) is characterized by the presence of all or a portion of the third chromosome 21. In this population, congenital heart disease (CHD) is the primary cause of death and morbidity in the first two years of life. Atrioventricular septal defect (AVSD) is the most common CHD seen in DS patients, followed by an atrial septal defect (ASD), ventricular septal defect (VSD), and tetralogy of Fallot (TOF). There is a high possibility of serious cardiac abnormalities in DS patients thus; there must be a protocol in place for early screening, prompt diagnosis and early operative measures. Due to advancements in technique and postoperative care over the last few years, the prognosis following surgical repair has substantially improved. Most of the post-operative strategies for patients with CHD can be applied to patients with DS. Physiotherapists play an important role in managing patients after the surgical repair of cardiac abnormalities. They also help in facilitating physical activity in children with DS. Our case report aims to provide the post-operative physiotherapy protocol for a child with DS operated for the closure of ASD and VSD along with the ligation of Patent Ductus Arteriosus (PDA). The main aim of post-operative cardiac and respiratory physiotherapy management was to clear lung secretions, promote healthy healing of the incision, maintain patency of lungs, reduce work of breathing and improve chest mobility while addressing delayed developmental milestones. The case report illustrates the importance of inpatient post-operative physiotherapy management which will lead to a gain in the existing knowledge while dealing with such patients.

**Keywords:** Congenital heart disease, Down syndrome, Trisomy 21, Atrial

septal defect, Ventricular septal defect, Cardiopulmonary Rehabilitation, physiotherapy, Vojta reflex therapy, Neurophysiological facilitation.

## 1. INTRODUCTION

When chromosome 21 fails to divide during gametogenesis, an extra copy of chromosome 21 remains in each body cell leading to trisomy 21. This genetic condition is called Down syndrome (DS). The majority of individuals with DS suffer developmental delays, mild to severe mental impairment and unusual facial features (Akhtar and Bokhari, 2022). Symptoms include mental impairment, early-onset vascular dementia and the formation of a variety of physical traits such as short stature, flattened nose and tiny, slanted eyes. They are also affected by additional health problems, such as thyroid problems, heart abnormalities and nutritional problems including obesity, hyperlipidemia and deficiency of vitamins and minerals (Mazurek and Wyka, 2015).

40 to 63.5% of all DS patients have congenital heart disease (CHD), which is the leading cause of death and disability in the early years of life (Benhaourech et al., 2016). The most frequent CHD found in these individuals is an atrioventricular septal defect (AVSD). Other CHDs include atrial septal defect (ASD), a ventricular septal defect (VSD) and a tetralogy of Fallot (Versacci et al., 2018). The basic structural trait of an AVSD is the existence of a shared atrioventricular junction (AVJ) as opposed to the discrete right and left AVJ in the normal heart. While in VSD, the defect is in the ventricular septum of the heart which results in a little amount of blood flowing into the right ventricle, leading to pulmonary hypertension (PH) (Asim et al., 2015).

Clinical examinations alone may not be able to identify the most frequent cardiovascular anomalies in new-borns with DS as their symptoms may be masked by the presence of persistently elevated pulmonary vascular resistance that is prevalent in these individuals (Versacci et al., 2018). Each healthcare team should be aware that these new-borns might have major heart conditions, and they should also have a screening process in place to guarantee early identification. The overall objective of advanced screening must be to recognize the infants with these defects early enough to make sure that by the time surgery can be implemented, almost none will already have irreversible pulmonary vascular changes (Dennis et al., 2010). A physical examination, electrocardiograph (ECG) and chest X-ray screening method were highly sensitive (95-99.8%) for identifying significant CHD in babies with DS. Universal neonatal echocardiography throughout the early years of life is advised by the most recent American Academy of Pediatrics (AAP) guidelines for health care for people with DS (Bogarapu et al., 2016).

Complete AVSDs are frequently surgically repaired during infancy. The common atrioventricular valve must be divided and resuspended to perform the traditional repairs for the complete AVSD by using either a single-patch technique or a two-patch technique in which the valve is not divided and individual patches are used to close each defect (Atz et al., 2011). The objective of timely closure is to lower the chance of early death or pulmonary vascular obstructive disease. Over the last several years, improvements in methods and postoperative care have improved the results of surgical repair. The age for elective repair is currently between three and six months in the majority of institutions, down from as recently as 1 year (St-Louis et al., 2014). We have presented the case of a 1-year-2-month-old female child with DS who underwent surgery for the closure of ASD and VSD with the ligation of PDA. The post-operative physiotherapy interventions were delivered in the inpatient setting with the primary aim to prevent post-operative complications and the secondary aim to promote neurodevelopmental milestones.

## 2. CASE PRESENTATION

### Patient Information

A 1-year-2-month-old female child, a known case of Down's syndrome with congenital heart disease was brought to the hospital with complaints of fever, cough and cold for 18 days. The child was all right 18 days back when she developed a high-grade intermittent fever of 103° F followed by cough and cold. Her parents took her to a pediatrician where she was managed with medications on an outpatient basis for 3 days. As the symptoms did not relieve after 3 days, she was admitted to a local hospital given an upper respiratory tract infection and treated with medications for 4 days. Then, the child was taken home in view of symptomatic relief after 4 days of hospitalization. The child developed similar symptoms with increased work of breathing in the next 2 days and was taken to another hospital for further management. She was admitted to Paediatric Intensive Care Unit (ICU) and was on oxygen support via a nasopharyngeal tube for 6 days. Parents were informed of a need for surgery for ASD and VSD closure and PDA ligation. After one month the child was brought for the same. The child underwent open surgery for ASD and VSD closure with PDA ligation.

As told by the mother, the child was all right till Day of Life 8 when she developed a fever, cough and cold. She was admitted to neonatal ICU in view of respiratory distress and managed further for 8 days. On day 21 karyotyping and 2-D ECHO were done.

Karyotyping suggested Trisomy 21 and 2-D ECHO findings were suggestive of ostium secundum ASD, VSD and PDA. Thus, the child was diagnosed with Down's syndrome with congenital heart disease. Since then, the child had frequent episodes of upper respiratory tract infection.

### Medical history

The patient is a known case of Down's syndrome with congenital heart disease and hypothyroidism. There is a history of admission to Neonatal ICU on day of life 8 for 8 days. Also, there is a history of 6 days pediatric ICU stay for 6 days one month back history of recurrent upper respiratory tract infection.

### Family History

History of second-degree consanguineous marriage. No other significant history.

### Developmental history

The history of gross motor, fine motor, social and language developmental history is summarised in Table 1.

**Table 1** History of developmental milestones

Milestones	The actual age of achievement	Expected age of achievement	Significance
Gross motor			
Neck holding	6 months	3 months	Delayed
Roll over	8 months	5 months	Delayed
Sitting with support	Yet to be achieved	6 months	Delayed
Fine motor			
Bidextrous reach	6 months	4 months	Delayed
Unidextrous reach	9 months	6 months	Delayed
Immature pincer grasp	12 months	9 months	Delayed
Mature pincer grasp	Yet to achieved	12 months	Delayed
Social			
Social smile	2 months	2 months	Normal
Recognizes mother	3 months	3 months	Normal
Stranger Anxiety	9 months	6 months	Delayed
Language			
Alertness to sound	1 month	1 month	Normal
Coos	3 months	3 months	Normal
Laugh loud	4 months	4 months	Normal
Monosyllables	6 months	6 months	Normal
Bisyllables	11 months	9 months	Delayed
Meaningful 1 to 2 words	Yet to be achieved	12 months	Delayed

### Immunization history

The child is immunized as per age.

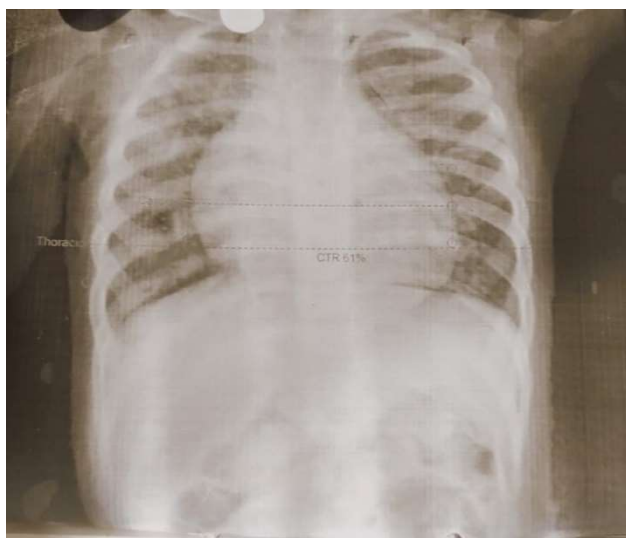
## 3. CLINICAL FINDINGS

The child was awake and alert. She was afebrile with a pulse rate of 88 beats per minute, respiratory rate of 32 breaths per minute, oxygen saturation of 100% on 4L O<sub>2</sub> via face mask and blood pressure of 110/56 mmHg. On observation, the patient was in a supine lying position. Flat face, epicanthic folds, brush field spots, tongue protrusion, short neck, small ears, small hands and feet and thick and taut skin were present. The child appears to have a weight and height lesser than expected for the given age. A median sternotomy scar was present on the chest. The child had abdominal breathing which was suggestive of the use of accessory muscles for breathing. The chest movements are bilaterally symmetrical. On palpation, apex beat is present in the left 5<sup>th</sup> intercostal space in

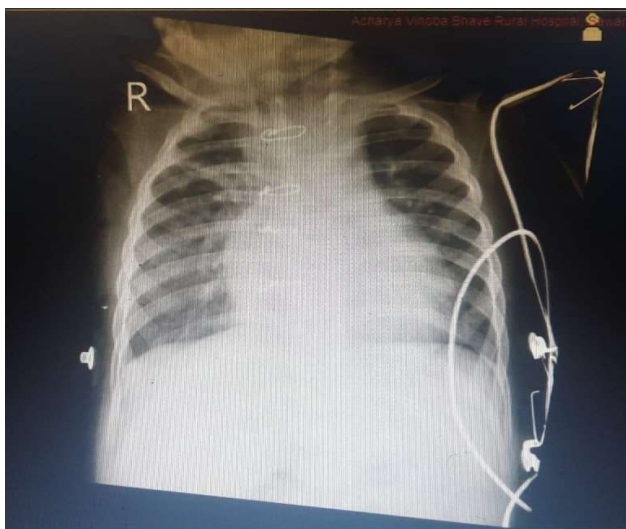
the midclavicular line, chest expansion is bilaterally symmetrical but reduced and the trachea is present centrally. On chest percussion, bilateral resonance is present, cardiac dullness's upper border was present in the left 2<sup>nd</sup> intercostal space, the right border is present at the right parasternal line and the left border is present at 2 cm left to the midclavicular line. On auscultation, air entry is bilaterally reduced in inframammary regions and no abnormal breath sounds and heart sounds were heard.

### Investigations

The Pre- and post-operative x-ray findings are given (Figure 1, 2).



**Figure 1** Pre-operative X-ray of posteroanterior view of chest suggestive of cardiomegaly and heterogenous opacities in bilateral lungs



**Figure 2** Post-operative X-ray of posteroanterior view of chest showing increased heart margins, increased broncho-vascular markings in right middle and lower zones, translucency in right upper zone and left upper, middle and lower zones

### Diagnosis

An operated case of ASD and VSD closure and PDA ligation.

### Treatment plan and interventions

The main aim of postoperative cardiac and respiratory physiotherapy management would be to clear lung secretions, promote healthy healing of the incision, maintain patency of lungs, reduce work of breathing and improve chest mobility. While addressing the cardiac and respiratory complications we also have to promote the development of the child. The physiotherapy interventions for the given case are as follows:

### *Nebulisation*

Apply nebulization with normal saline at the start of each session.

### *Postural drainage with Manual chest percussions and vibrations*

Perform percussions and vibrations for 2 minutes on each segment as given (Figure 3, 4). The frequency would be twice a day daily. For drainage of apical segments, the head end of the child would be inclined to 45 degrees. Perform percussions and vibrations on each side of the chest from the midclavicular line to the shoulder.

To drain posterior segments, the child would be leaning forward on the pillow. Perform percussions and vibrations on the scapular region on each side.

To drain the middle lobe and lingula segment, the child would be in a side-lying position and roll back quarter turn on both sides. Perform percussions and vibrations over the nipple area on each side with the child in respective positions.

To drain lateral basal segments, the child would be in a side-lying position on each side. Perform percussions and vibrations over axillary areas on each side with the child in respective positions. Avoid percussions and vibrations over the lower ribs.

To drain anterior basal segments, the child would be supine lying position. Perform percussions and vibrations over the upper margin of the ribs on each side. Avoid percussions and vibrations over the lower ribs and stomach.

Drainage of posterior basal segments would start at the end of the first-week post-surgery as the child has to be taken into a prone lying position and that would put undue pressure on the incision site. Perform percussions and vibrations on the lower ribs in the same position.



**Figure 3** Application of percussion with the child in side lying position.

### *Cough stimulation*

It is performed after each cycle of chest percussion and vibrations. Tracheal stimulation is used to stimulate cough. Place a finger just above the sternal notch to apply gently firm pressure circularly during the expiration. This helps to expel the secretions from the airways.

### *Suctioning*

Oral and nasal suctioning is performed after each session of chest percussion and vibrations.



***Vojta reflex rolling therapy***

Helps in increasing the depth of coastal respiration with an expansion of the lung. Positioning the child in supine lying with legs flexed to 45 degrees. 5 minutes of stimulation of the pectoral area on each side.



**Figure 4** Application of vibrations using index and middle finger with the child in side lying position

***Neurophysiological facilitation of respiration (NFR)***

Apply the techniques after 10 days as it gives time for sutures to heal. The child would be in a supine lying position with arms by the side. Application of inter costal stretch with index and middle finger over 2<sup>nd</sup> and 3<sup>rd</sup> ribs during exhalation for 2 minutes. After the rest of 1 minute, the second technique would be vertebral pressure in which firm gentle pressure is applied to the upper and lower thoracic vertebrae to improve epigastric excursion and apical thoracic inspiratory movement respectively for 2 minutes each.

***Lap therapy***

With the therapist in long sitting with his back rested against the wall have the child seated on the lap with support to facilitate the development of sitting without support. At the same time give some small colourful objects to the child to develop a mature pincer grasp. Teach the same to the mother and ask her to perform it several times a day.

After 2 weeks of management, the patient was discharged. The exercises and interventions to be performed at home were taught to the parents before the discharge.

***Follow up and outcomes***

After two weeks of physiotherapy management of the patient, the outcomes were assessed and compared with pre-interventional scores. The patient showed improvements in cardiorespiratory parameters, Faces Pain Scale and Paediatric Respiratory Severity Score. The comparison of pre-and post-interventional scores is given (Table 2).

**Table 2** Comparison of pre and post outcome measure scores

Sr. no.	Outcome measure	Pre-intervention	Post-intervention
1	Blood pressure	110/56 mmHg	95/60 mmHg
2	Heart rate	140 beats per minute	100 beats per minute
3	Oxygen saturation	91% to 93 %	98 % to 100 %
4	Respiratory rate	50 breathes per minute	35 breathes per minute
5	Respiratory pattern	Irregular abdominal breathing with the use of accessory muscles	Regular Thoraco- abdominal breathing without the use of accessory muscles
6	Chest expansion at nipple level	1 cm	3 cm
7	Faces pain scale	8 (hurts a lot)	2 (hurts a little bit)
8	Paediatric Respiratory Severity Score	4/5 (severe)	1/5 (mild)

#### 4. DISCUSSION

Many of the perioperative assessment and care guidelines for CHD patients also apply to those with DS. The risk of pulmonary hypertension must be carefully considered when considering cardiac surgery and both cardiac and non-cardiac disorders must be evaluated (Delany et al., 2021). Infants seeking surgical treatment for CHD have the possibility of acquiring neurodevelopmental disorders, which may not show up until months after being discharged from the hospital. Thus, postoperative gross motor deficits bother parents of infants with CHD in the majority of cases. Early surgical effects in children with CHD that result in a gross motor developmental delay persist for a very long time, if not treated therapeutically. Exercises for the extremities and trunk are used in physical therapy to help patients' gross motor skills. These exercises are performed one to three times a day for 20 to 60 minutes each, five or six days a week, to prevent respiratory issues and to help patients. Postural drainage treatment, thoracic expansion, chest mobilization and squeezing are all parts of respiratory physical therapy (manual breathing assists with postural drainage) (Haseba et al., 2018).

The role of a physiotherapist while working treating a patient with Down syndrome varies according to the individual's life stage and often includes promoting physical exercise. Early in childhood, the emphasis is on enhancing motor abilities and preventing the development of maladaptive compensatory movement patterns, depending on an individual's presentation (Shields, 2021). We have presented the case of a 1-year-2 months-old child with DS who underwent an operation for the repair of a congenital heart defect. The patient also has delayed developmental milestones. The primary goal of postoperative physiotherapy management was to clear the secretions and improve ventilation of the lungs along with the secondary goal of facilitating of normal development of the child.

Chest physiotherapy (CPT) has been suggested as one of the strategies that can aid in the clearance of extra tracheobronchial secretions, which assist in improving ventilation. CPT consists of various techniques, involving postural drainage and positioning and active techniques such as percussion vibrations and suction. Positioning is an important part of respiratory physiotherapy, yet its importance is often undervalued. Despite the preference for prone over supine and side-lying as the recommended posture for enhanced oxygen uptake, quarter turning from prone offers the potential to maintain the benefits of prone while limiting its drawbacks (Hough, 2009). We had recommended the parents keep the child in a quarter turn from a prone position for most of the time in the early postoperative phase. This allowed improvements in oxygenation and ventilation. As the sutures over the chest wall healed, we then suggested the prone lying position.

Vojta therapy is a physical therapy technique that has been shown effective in treating adults and children with respiratory disorders. The technique involves the application of bearable pressure to specific aspects of the patient's body to reflexively engage the ventilatory physiology (Lopez, 2021). Chest physiotherapy (CPT) helps in clearing secretions, preventing atelectasis after extubation and lowering the need for re-intubation and thereby, helping in improving lung ventilation. The Vojta method is a form of physical therapy that uses isometric strengthening with tactile stimulation to promote the emergence of healthy movement patterns and, as a result, enhance breathing (Giannantonio et al., 2010).

Neurophysiological facilitation of breathing involves the application of external proprioceptive and tactile stimuli to produce reflex respiratory movement responses that aid in changing the pace and depth of respiration. It includes intercostal stretching, vertebral pressure, anterior stretching and basal lift. This approach facilitates the activation of respiratory muscles and improves breathing pattern, chest expansion, epigastric excursion and pressure over the vertebrae, which increases respiratory movement (Patel and Prajapati, 2019). The use of Vojta therapy and neurophysiological facilitation of breathing helped in preventing

respiratory complications after the surgery. At the same time, it helped in addressing the neurodevelopmental concerns related to DS.

## 5. CONCLUSION

Two weeks of physiotherapy management for a child with DS who had undergone open heart surgery for the repair of ASD and VSD was aimed at preventing post-operative complications. Interventions to clear airways, improve chest expansion, reduce work of breathing, improve respiratory muscle activity and facilitate the development of the child were implemented in the in-patient department. There were improvements in respiratory and cardiac parameters following two weeks of post-operative physiotherapy management. Thus, we conclude that applied physical therapy interventions are effective in the post-operative (ASD and VSD closure with PDA ligation) care of a 1-year-old child with Down's syndrome.

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## Author Contributions

AD, DC, AG, VT and VV: Contributed to the design and implementation of the research and to the writing of the manuscript.

## Informed consent

Written & Oral informed consent was obtained from participant's parents.

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## Conflict of interest

The authors declare that there is no conflict of interests.

## Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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