

Pediatric Oro-Pharyngeal Dysphagia Secondary to Post Diphtheric Bulbar Palsy with Dilated Cardiomyopathy

Shalini S Narayanan*

Swami Rama Himalayan University, Department of Ent, Department of Ent Department of Ent, India.

***Corresponding Author:** Shalini S Narayanan, Swami Rama Himalayan University, Department of Ent, Department of Ent Department of Ent, India.

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Abstract:

Diphtheria happens to be a serious bacterial infection which adversely affects the mucous membranes of the nose and throat. It is caused by *Corynebacterium diphtheriae* which multiplies on or near the surface of the throat. It can also multiply on the skin and this usually spreads through airborne droplets or contaminated household and personnel belongings.

Key words: quadriparesis; hypotonia; dysphonia; dysphagia; nasogastric tube

Introduction

Diphtheria happens to be a serious bacterial infection which adversely affects the mucous membranes of the nose and throat [1]. It is caused by *Corynebacterium diphtheriae* which multiplies on or near the surface of the throat. It can also multiply on the skin and this usually spreads through airborne droplets or contaminated household and personnel belongings [2]. Diphtheria can be treated with medication in the initial stages but in the advanced condition it can lead to breathing problems, heart damage and nerve damage. Studies have shown that the incidence of cardiomyopathy following diphtheria is 10-20% [3][4]. The clinical features of diphtheritic cardiomyopathy become evident by the 2nd week of infection and in severe cases may present with most features [4]. Cardiomyopathy is referred to the condition where the myocardium i.e., the heart muscles get affected. Dilated cardiomyopathy is the condition where the heart chambers get dilated [5]. Pediatric cardiomyopathy can be seen to affect children and teenagers of any gender, race or age. It can even be more common in infants than in older children. It has been documented that some children may show no cardiomyopathy symptoms till the time they experience a sudden cardiac arrest [6]. Severe complications of diphtheria have highlighted polyneuropathy with bulbar, respiratory as well as circulatory disturbances. Cranial nerves have also been affected thereby showing signs of quadriparesis, hypotonia, dysphonia, dysphagia and other complications [7].

Case Report:

A child aged 11 years/ male was initially asymptomatic but then he developed pain in the abdomen which was sudden onset, mild to moderate in intensity, generalized and continuous. The pain was relieved temporarily on taking medications. There was complaint of acute

breathlessness from the last 4 days which was progressive in nature. The child also developed progressive and productive cough. The child was then brought to the emergency of HIMS for breathing support. Initial treatment was provided, once the condition was slightly stable the child was shifted to ICU. Pleural tapping and infusion dopamine was started. On the 8th day the child was shifted to pediatric ward. After that the child was referred to the Rehabilitation Unit of Speech & Hearing, Department of ENT after a month with the complaint of palatal weakness and change in voice. Laryngeal and oral cavity endoscopy revealed reduced mobility of the soft palate and paresis of vocal folds with the presence of phonatory gap. The child was on Nasogastric tube for 39 days due to respiratory distress caused due to pooling of secretions.

Detailed speech and language evaluation was carried out by the Speech Language Pathologist focusing the domains of speech and swallowing. Consent was taken from the child's caregiver. Tools used for assessment included Milestones for Early Communication Development (MECD) for language evaluation, GRBAS for voice evaluation, Gugging Swallowing Screen (GUSS) and The Dysphagia Disorder Survey (DDS) for swallowing evaluation and AYNJIHH Intelligibility rating scale for intelligibility of speech [8][9][10][11].

The child was co-operative during the entire assessment procedure. The assessment included Oral Peripheral Mechanism Examination which showed reduced movement in the soft palate during the production of /a/ leading to a perceptual nasal emission. Fog test was done to confirm the nasal emission. Language assessment showed adequate receptive and expressive language skills in the segments of Semantics, Syntax, Morphology and Pragmatics. GRBAS was administered and was indicative of mild hoarse voice. The intelligibility rating score suggested

that the speech was understood with concentration and effort especially by sympathetic listener.

Dry swallow test was done which showed delayed laryngeal elevation. GUSS suggested severe dysphagia with high risk of aspiration. NPO was advised and the caregiver was counseled about the risk of aspiration. DDS was carried out in two parts- Part 1 related factors showed poor Body Mass Index, Good independence and body postural control. Part 2 consisted of Feeding and Swallowing Competency. The child was on NG tube with the advice of NPO.

Accumulation of all the evidences led to the diagnosis of oro-pharyngeal dysphagia with palatal weakness and mild hoarse voice secondary to bulbar palsy. The child was given regular follow ups for speech and swallow therapy sessions. The child was demonstrated swallowing maneuvers – effortful swallow and super supraglottic swallow. Cold sour stimulation was given to elicit swallowing reflex. Oromotor exercises and

soft palate elevation was worked on. Unilateral digital pressure was given for adduction of vocal folds.

After 2 sessions of speech and swallow therapy soft solids and thick liquid was introduced. The food was given in 3ml quantity and the child was asked to follow the maneuver with food. Swallow was effortful but it was complete and there was no oral residue, no involuntary cough as well as no gurgly voice after the swallow. After 4 sessions of speech and swallow therapy the NG tube was removed and the child was introduced to thin liquids in sips with chin tuck. Post therapy evaluation was done after 5 sessions in which GRBAS showed clinically normal voice. The intelligibility rating scale showed a score of 2 and GUSS score indicated slight dysphagia with low risk of aspiration. Dysphagia Disorder Survey –Part 1 showed poor body mass index, good independence and body postural control, soft solids were recommended in spoonful with swallowing maneuvers.

	PRE- THERAPY			POST THERAPY		
	Chewable solid	Non chewable solid	Liquid	Chewable solid	Non chewable solid	Liquid
Orientation	0	0	0	0	0	0
Reception	0	0	0	0	0	0
Containment	0	0	0	0	0	0
Oral transport	1	1	1	0	0	0
Chewing	0	-	-	0	-	-
Oral-pharyngeal swallow	1	1	1	0	0	0
Post swallow	1	1	1	0	0	1

Table 1: Feeding and Swallowing Competency-Dysphagia Disorder Survey (comparison pre and post swallow therapy)

SI No	Tests Administered	PRE-THERAPY			POST THERAPY		
		Scores Obtained	Maximum Score	Interpretation	Scores Obtained	Maximum Score	Interpretation
1	GRBAS	G-1 R-1 B-1 A-0 S-0	0	Mild Hoarse Voice	G-0 R-0 B-0 A-0 S-0	0	Clinically normal voice
2	GUSS	4	20	Severe dysphagia	19	20	Slight dysphagia with low risk of aspiration
3	AYJNIHH Intelligibility rating scale	3	0	Speech understood with concentration and effort especially by sympathetic listener	2	0	Speech can be understood with little effort. Occasionally needs repetition.

Table 2: Formal test results –pre and post speech therapy

The child was discharged with recommendation for soft solids and cautious thin liquids. With an improvement in his overall condition, he was discharged from the hospital after 48 days accompanying a follow up protocol.

Discussion:

The clinical features of diphtheritic cardiomyopathy become evident by the 2nd week of infection and in severe cases may present with most

features [12]. In India most people with diphtheria are either partially immunized or unimmunized. In our case the immunization details were not available. Presence of myocarditis has been reported in 16.5% in a retrospective case series from India [13]. In a study done on 154 Vietnamese children with the clinical diagnosis of diphtheria 13 children had diphtheritic cardiomyopathy during the time of hospital admission and 19 children had developed subsequently [12]. 8 children had even developed diphtheritic neuropathy as a late complication. Hoarseness of voice with difficulty in managing secretions were also commonly seen

these children. Neuropathy develops within three to five weeks of diphtheria with the initial onset of bulbar palsy followed by cranial nerve involvement. In this case profile there was presence of palatal paresis as well as phonatory gap leading to hoarseness of voice. Along with these, presence of severe Oro-pharyngeal dysphagia confirmed Cranial nerves IX and X being affected. These findings were corroborated in the study by Piradov [7]. Another study showed the presence of bulbar palsy in all 13 with isolated palatal palsy in 7 children and progressive cranial nerve involvement was seen in 6 children leading to quadriplegia [14]. There is a dearth of literature which highlights the speech and swallowing profile specifically due to bulbar palsy in diphtheritic neuropathy. Children with diphtheritic complications are challenging to manage. Often, several organ systems are affected by the infection and toxin. However, early identification would definitely help to manage the speech and swallowing complications symptomatically. It is important to focus on motivating targets. In this case working on oromotor function and elicitation of swallow reflex thereby aiding in oral feed increased the motivation in the child. Swallowing can definitely be one of most important function that can stand to uplift the overall quality of life and further promote working on other vital functions. Disease prevention through vaccination and early identification and intervention stands to be the vital keys in reducing the morbidity and mortality of this particular disease.

Conclusion:

Pediatric dysphagia is challenging for SLPs to evaluate. A descriptive assessment should be done to identify all the overlapping conditions which include both clinical and instrumental evaluation tools. Aspiration risk can always be confirmed by the use of instrumental assessment which was not available in the present set up [15]. Prompt treatment is important for specific management of symptoms contributing to speech and swallowing disorder. This case profile will help professionals working in neurorehabilitation unit to expand the horizon of understanding with the assessment and management of challenging profiles.

Compliance with Ethical Standards:

Conflict of interest- The authors have no conflict of interest.

Ethical approval: All procedures performed in the study were in accordance with the ethical standards of the institutional research committee.

Informed Consent: Written informed consent was obtained from the family members of the participant included in the study.

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