

Original Research Paper

## Neurological Etiologies Necessitating Elective Tracheostomy in Pediatric Age Group: A 100 Patient Experience

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### ABSTRACT:

**Background:** Tracheostomy is a life-saving procedure and is indicated in a variety of peripheral or central conditions leading to respiratory compromise. Mainly, tracheostomy is done for three reasons; to bypass obstructed upper airway, to remove secretions and keep the airway patent; and to effectively ventilate the patient. **Objective:** The aim of this study is to quantify the frequency of different neurological disorders which indicate elective tracheostomy to avoid respiratory compromise and its associated complications. **Materials and Methods:** In this Prospective, observational study, 100 pediatric patients with confirmed neurological diagnosis and associated severe respiratory impairment requiring invasive mechanical ventilation were enrolled. The study was conducted over the period of 3 months, from May 2022 to July 2022, at Children Hospital and Institute of Child Health, Lahore. **Results:** According to the collected data, prevalence of male patients was almost twice as that of females (2.9:1). The main indication of tracheostomy was the requirement of prolonged mechanical ventilation due to neurological condition (n= 36, p<0.02). Among all the neurological etiologies, the CNS infections with serious respiratory manifestations were most common including Guillain Barre Syndrome (n=26, p=<0.01), Diphtheria Polyneuropathy (n=8, p=0.06), Hypoxic-Ischemic Encephalopathy (n=5, p<0.08), Varicella Zoster Infection (n=3, p=0.09), Poliomyelitis / Post-Polio Syndrome (n=2, p<0.1), and Tetanus (n=1, p=0.15). **Conclusion:** Infectious neurological diseases such as GBS, Diphtheria, and polio, causing respiratory failure are still prevalent in developing countries like Pakistan & India and are the main causes of tracheostomy in pediatric age group. Prolonged mechanical ventilation was the main indication for tracheostomy.

**Keywords:** tracheostomy, pediatrics, infections, GBS, ETT, neurological intensive care

### INTRODUCTION:

Tracheostomy is one of the ancient surgical procedures to bypass upper airway tract obstructions. About 40 years ago, pediatric tracheostomy was only indicated in the cases of infectious diseases of upper airway such as

Diphtheria, Epiglottitis etc. [1]. Over the time, the indications and applications of tracheostomy were revised and evolved, considerably. With the administration of vaccines, the incidence of infectious diseases caused by *Corynebacterium Diphtheria* and

*Hemophilus Influenzae* has decreased dramatically [2]. Hence, the number of pediatric emergency tracheostomies has fallen, globally. However, with the introduction of other indications of tracheostomy, the overall number of tracheostomies has increased over time [3]. Worldwide, the pediatric tracheostomy is performed in tertiary care facilities only, in children with acute or chronic, congenital, or acquired respiratory and/or neurological impairment, and in children with long term dependency on invasive mechanical ventilation [4]. There are a number of neurological etiologies that can result in severe respiratory impairments. Emergent Endotracheal Tube Placement with mechanical ventilation can save the patient temporarily but if the long term invasive mechanical ventilation is required, ETT must be replaced by tracheostomy [5]. According to the experts, if patient needs ETT and mechanical ventilation for 7 days or more, then tracheostomy should be performed for sustainable and secured airway [6]. Although in some cases of severe neurological impairment, patients need ETT assisted mechanical ventilation for a longer period of time, for example in case of a brain injury with poor GCS and unconsciousness, tracheostomy cannot protect the airway against secretions and gastric aspiration as ETT does [7]. Tracheostomy decreases the work required for breathing but that is not an issue for the patients with brain injury. Prolonged ETT intubation is associated with high risk of nosocomial pneumonia and prolonged ventilator dependency which can be significantly reduced by tracheostomy. Prolonged ETT intubation is also associated with a number of tracheal complications especially in the patients with severe brain trauma. If patient is intubated for more than 14 days, there is a high risk of development of serious complications such as laryngeal injury including laryngeal edema and inflammation, vocal cord injury, ulceration, paresis, paralysis, granuloma, and stenosis. So, it is important to weigh the pros and cons after regular and careful clinical evaluations of the patients [8]. There are a variety of neurological disorders which require tracheostomy as a part of an active or long-term management plan. These diseases/disorders include congenital/chromosomal/neurometabolic, idiopathic, traumatic, infective, autoimmune, and inflammatory disorders [9]. The incidence of infective neurological causes has decreased worldwide over the time, by the

virtue of effective immunization. Nevertheless, these are still prevalent in developing countries due to poor vaccination coverage and healthcare provision. There are several neurological infections that cause acute flaccid paralysis and hence the respiratory compromise. The mechanism of respiratory disability may vary slightly, but the ones that are mentioned in the article required tracheostomy at some point in the management of the disease [9, 10]. Among all the acute neurological infections, Guillian Barre Syndrome (GBS) is the most common cause of peripheral, motor neuropathy with presumed antecedent infection and autoimmune etiology. Usually, the infection caused by *Campylobacter Jejuni* precedes the GBS with rapidly progressive axonal injury, ascending paralysis, respiratory failure, severe residual disability and slow recovery [11]. Globally, the incidence of GBS ranges from 1.1 to 1.8 per 100,000 people, annually [12]. The overall incidence of GBS increases with the age; peaks after 50 years. In Pakistan, the exact incidence and prevalence is not known. Electrophysiological studies for early detection, definitive diagnosis, and classification of GBS play an important role in significant reduction of morbidity and disability [13]. Poliomyelitis or Post Polio Syndrome is caused by highly infective, Enterovirus via feco-oral route [14]. It is a type of acute flaccid paralysis associated with prolonged or permanent flaccid muscle paralysis, respiratory failure, and death. Due to combined efforts, this sporadic, highly contagious infection is successfully eradicated from the world, except some areas of Sub-Saharan Africa and South Asia where it is still an endemic. In 95% of the infectious cases, the patients either remain asymptomatic or have a flu-like illness [15]. Despite effective preventive and management policies, some patients develop late functional complications after a long period of complete stability and recovery. This condition is called Post-Polio Syndrome. The bulbar form of Poliomyelitis involves high morbidity and mortality due to vasomotor, circulatory, and autonomic dysfunction along with respiratory failure. The acute respiratory failure needs emergent intubation and prolonged mechanical ventilation which can later indicate elective tracheostomy [16]. Diphtheritic Polyneuropathy (DP) is a serious bulbar, respiratory, and circulatory complication. It is a direct and acute indication for

intubation and tracheostomy [17]. The etiology causes a decrease in lung's vital capacity (<16mL/kg body weight), paralytic collapse of laryngeal muscles, and multiple cranial nerves impairment causing peripheral motor disturbances. Diphtheritic Polyneuropathy is also a late complication of diphtheria infection, so special attention should be given from the fourth to seventh week of DP [18]. The objective of this study is to determine the current neurological indications of tracheostomy in the pediatric age group and to assess the burden of infectious causes with central involvement in developing countries like Pakistan, despite the discovery and implementation of effective immunization all around the globe.

**MATERIAL & METHODS:**

In this prospective, observational study, 100 patients were recruited by following the inclusion and exclusion criteria. The study was conducted over the period of 3 months, from May 2022 to July 2022 at a tertiary care hospital. The Qualitative and Quantitative data was collected for 100 selected patients who were registered through the CH & ICH, Lahore. Multi-departmental Collaboration was ensured. Verbally explained, written informed consent was taken from the guardians, mostly from the parents of the children. The whole purpose and procedure of this study was explained to the attendants. Few parents refused to take part in the study, their rights were respected, and they were not included in our study. By any means, this study does not adversely affect the rights and welfare of the subjects. All the patients had some neurological manifestation as a primary cause for elective tracheostomy. Those patients were also not included in the study which needed tracheostomy due to

some other primary cause while the neurological condition co-existed.

**INCLUSION CRITERIA:**

- Patients diagnosed with neurological impairments that were compromising the respiration by any means, i.e. Upper airway obstruction, Vocal cord paralysis/paresis, respiratory muscle paralysis/paresis, altered sensorium, skeletal/laryngeal muscle weakness, prolonged mechanical ventilation, etc.
- Patients who were on mechanical ventilation for > 7 days
- Patients who required prolonged mechanical ventilation or on whom, ventilator weaning off attempts were performed but failed

**EXCLUSION CRITERIA:**

- Patients of age more than 14 years
- Patients who needed tracheostomy for causes other than neurological anomaly
- Patients with incomplete previous medical record
- Patients with underlying diseases (other than neurological issues) such as hypothyroidism, diabetes, hepatic or renal dysfunction, vasculitis, metastasis, intoxication, etc.

**RESULTS:**

According to the collected data, there were 67 males (p <0.02) and 23 females (<0.04) with male to female ratio of 2.9:1. The peak incidence of tracheostomy is seen at the ages of 2-5 years (n=29, p <0.05) and 6-12 months (n=21, p<0.05). P value less than 0.05 is considered significant. The demographic characteristics including age group and gender are described in Table 1.

<b>Demographic Characteristics</b>		<b>Frequency (%age)</b>	<b>P-value</b>
<b>Age</b>	0-6 months	16 (16%)	Not Significant
	6-12 months	21 (21%)	<0.05
	1-2 years	11 (11%)	Not Significant
	2-5 years	29 (29%)	<0.05
	5-10 years	10 (10%)	Not Significant
	10-14 years	13 (13%)	Not Significant
<b>Gender</b>	Male	67 (67%)	<0.02
	Female	23 (23%)	<0.04

**Table 1 Demographic Characteristics of selected children (n=100)**

Eight different indications of elective tracheostomy in pediatric population were documented in this study. In descending order, the 3 main indications are prolonged invasive mechanical ventilation (n=36, p=<0.02), Upper

airway obstruction (n=20, p=<0.04), and Metabolic/ Genetic/ Chromosomal anomalies (n=16. p=<0.05).

Indications of Tracheostomy	Frequency (%age)	P-value
Upper Airway Obstruction	20 (20%)	<0.04
Prolonged Invasive Mechanical Ventilation	36 (36%)	<0.02
Hypoxic Brain Injury	8 (8%)	Not Significant
Metabolic / Genetic / Chromosomal	16 (16%)	<0.05
Cerebral Hemorrhage	4 (4%)	Not Significant
CNS Infection	9 (9%)	Not Significant
Demyelinating Disorder	5 (5%)	Not Significant
Brain Tumor	2 (2%)	Not Significant

**Table 2 Indications of Elective Tracheostomy in children with neurological disease associated respiratory compromise**

Among the 100 selected patients, 16 different neurological conditions were documented, all leading to the respiratory failure by different mechanisms. Surprisingly, 45% of the total patients had neurological manifestation due to infectious etiology. The infectious causes of neurological conditions include (in descending

order) Guillain Barre Syndrome (n=26, p=<0.01), Diphtheria Polyneuropathy (n=8, p=0.06), Hypoxic-Ischemic Encephalopathy (n=5, p<0.08), Varicella Zoster Infection (n=3, p=0.09), Poliomyelitis / Post-Polio Syndrome (n=2, p<0.1), and Tetanus (n=1, p=0.15).

Neurological Conditions	Frequency (%age)	P- value
Poliomyelitis / Post-Polio Syndrome	2 (2%)	Not Significant
Guillain Barre Syndrome	26 (26%)	<0.01
Myasthenia Gravis	14 (14%)	<0.03
Diphtheria Polyneuropathy	8 (8%)	Not Significant
Varicella Zoster Infection	3 (3%)	Not Significant
Duchenne Muscular Dystrophy	5 (5%)	Not Significant
Hypoxic-Ischemic Encephalopathy	5 (5%)	Not Significant
Cerebral Palsy	8 (8%)	<0.05
Spinal Muscular Atrophy (SMA)	3 (3%)	Not Significant
Neurofibromatosis	4 (4%)	Not Significant
Brain Tumors	6 (6%)	Not Significant
Kernicterus	5 (5%)	Not Significant
Leukodystrophy	2 (2%)	Not Significant
Amyotrophic Lateral Sclerosis	3 (3%)	Not Significant
Traumatic Brain Injury	5 (5%)	Not Significant
Tetanus	1 (1%)	Not Significant

**Table 3 Spectrum and frequency of various neurological conditions necessitating elective tracheostomy**

During the period of 3 months of this study, no tracheostomy related mortality was documented. None of the patients underwent decannulation. The limitations of this study are that the duration of three months period did not allow the proper follow up of the patient and the prognosis, duration, and late complications of tracheostomy could not be assessed.

**DISCUSSION:**

Tracheostomy is an operative procedure that creates a surgical opening on the anterior aspect of cervical trachea, mainly for effective ventilation [1, 2]. Patients

with tracheostomy are a heterogeneous group of population with various indications and associated conditions. In this study, we only included the patients from pediatric age group requiring elective tracheostomy due to any congenital or acquired neurological condition [3, 4]. Children with congenital or acquired neurological disorders are prone to have respiratory co-morbidities [5]. There are multiple neurological manifestations that cause respiratory compromise such as pharyngeal/laryngeal muscle weakness leading to Vocal cords dysfunction, respiratory muscle weakness or paralysis causing hypoventilation, skeletal muscle weakness i.e.

SMA associated with prolonged immobility, pooling of respiratory secretions in respiratory tract, recurrent lung infections, and irreversible destruction of lung parenchyma [6, 7]. In patients with Cerebral Palsy and other neuromuscular disorders, pharyngeal and laryngeal muscles fail to maintain the patency of upper airway tract against inspiratory negative pressure [8, 9]. The hypotonic pharynx tends to remain collapsed causing upper airway narrowing or obstruction. Whenever, the patient develops acute upper or lower respiratory tract infection, the condition worsens with additional mucosal swelling and pooling of respiratory secretions due to ineffective clearing of sputum [10]. Moreover, the muscular weakness also results in disrupted swallowing mechanism causing GERD (Gastro-Esophageal Reflux Disease) and micro aspirations due to prolonged immobility and loss of protective cough reflex. All these factors make the patient prone to develop permanent lung damage via recurrent bronchitis, bronchiectasis, and pneumonia [11].

Tracheostomy has several therapeutic as well as palliative applications in patients with neurological diseases. As discussed in this article, tracheostomy provides secure and sustainable pathway for short or long-term invasive mechanical ventilation. It secures the breathing pathway in case of upper respiratory tract obstruction. It reduces the physiological dead space and significantly reduces the work required for breathing [12]. It provides effective, less traumatic and easily accessible route for airway toileting via sputum suctioning which is essential, especially during acute respiratory tract infections. In the case of emergency positive pressure ventilation, tracheostomy provides a quick, accessible route and significantly decreases the morbidity and mortality caused by delayed or unsuccessful attempts of endotracheal tube placement. Although noninvasive ventilation is an alternative emergency maneuver, but in the patients with neurological diseases, the invasive ventilation is preferred due to the high risks of secretions and gastric content aspiration. It also provides a safe route for diagnostic as well as therapeutic endoscopy, reduces the duration of procedure, and significantly minimizes the procedure associated risks [13, 14]. In most of the conditions, early tracheostomy proves to be highly beneficial in terms of decreasing morbidity and disability [23]. For example, in GBS, early tracheostomy

can end the dependency on mechanical ventilator, providing more comfort and a chance of commencement of early oral nutrition with adequate oral hygiene that can speed up the weaning process [24]. It also adds to the parent's satisfaction and patient's motivation for the quick recovery. But the decision of tracheostomy is often critical, and it needs expert's opinion and evaluation to assess the pros and cons of tracheostomy. In the neurological cases where there is an increased risk of aspiration, ETT intubation with inflated cuff provides more protection and safer airway. However, ETT is not a sustainable way to maintain airway and is associated with various early and late complications [25-27]. In the literature, the indications of tracheostomy are not found consistent throughout the world. In developed countries, tracheostomy is mainly indicated in traumatic, chromosomal, autoimmune, or inflammatory conditions [15-18]. Whereas, in developing countries, the major causes requiring tracheostomy are infections and their peripheral and central ramifications. In Pakistan, the vaccination rate in children is considerably lower than that of developing countries due to ineffective surveillance and lack of general public awareness [19, 20]. However, the unvaccinated or partially vaccinated are not the only ones susceptible to infectious diseases. The severe neurological infectious diseases such as GBS, Diphtheria etc., are also found common in fully immunized children. In that case, there are multiple reasons to consider, but the experts speculate that the main reason for this poor coverage is low quality vaccination. In developing countries like Pakistan, less immunogenic vaccinations are prevalent because of limited resources and cost effectiveness [21, 22]

## **CONCLUSION**

This study demonstrates the frequency of different neurological conditions with significant respiratory complications that necessitate surgical tracheostomy. All over the world, the infectious causes of tracheostomy have almost diminished but these are still prevalent in our setup. According to this study, prolonged mechanical ventilation was the most common indication and GBS was the most common neurological condition requiring elective tracheostomy for better prognosis in children.

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