

Table 1: Baseline Demographic and Clinical Characteristics

Patient Characteristics (n=622)	Median (IQR) or N (%)
Median age of NIV Initiation (years)	7.8 (9.2)
Males	378 (60.8)
Primary Underlying Diagnostic Category	
Upper Airway	371 (60)
Central Nervous System	107 (17)
Musculoskeletal	93 (15)
Cardiopulmonary	39 (6)
Other	12 (2)
Number of comorbidities	
0	50 (8)
1-2	310 (50)
3-4	161 (26)
5 or more	101 (16)
Additional Technology Used	
Daytime oxygen	30 (5)
VP shunt	17 (3)

G-tube/NG tube feeding	99 (16)
Wheelchair use	63 (10)
Prior Upper Airway Surgery	293 (47)
Prior Other Major Surgery	231 (37)

Table 2: Breakdown of diagnostic categories and underlying disease conditions of children started on long-term non-invasive ventilation.

Diagnostic Category (%)	Underlying Disease Conditions
Upper Airway (60%)	<ul style="list-style-type: none">- Upper airway narrowing/malformations (narrowing of subglottic/supraglottic cleft, airway malacias, Pierre Robin sequence, other airway abnormalities)- Down syndrome- Prader Willi- Obesity, Diabetes or Metabolic Syndrome
Central Nervous System (17%)	<ul style="list-style-type: none">- Congenital or acquired brain injury- Autism spectrum disorders- Central hypoventilation syndromes
Musculoskeletal (15%)	<ul style="list-style-type: none">- Spinal Muscular Atrophy Types 1,2 and 3- Duchenne Muscular Dystrophy- Other muscular dystrophies- Myopathies- Achondroplasia
Cardiopulmonary (6%)	<ul style="list-style-type: none">- Congenital heart disease or cardiac failure- Cystic fibrosis, interstitial lung diseases and other chronic lung diseases- Bronchopulmonary dysplasia or chronic lung disease of prematurity

Other (2%)	<ul style="list-style-type: none">- BPD/CHD and myopathy- Diagnosis not available
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Table 3: Univariate Analysis of Demographic, Clinical, Technology-Related, and Follow-up Variables for Children Using Long-term Non-invasive Ventilation. Variables with a $p < 0.20$ on univariate analysis were included in the regression models. The following variables were forced into all models due to clinical relevance: Age, sex, disease category, number of comorbidities, number of additional technologies, NIV type. Bold indicates $p < 0.05$

	Outcomes with p-values					
Co-variates	Continuation on NIV	Discontinuation due to improvements in underlying condition	Patient/Family NIV Declination	Switch to IMV or Death	Transfer to other services	Hospital Admissions
Age	0.008	<0.001	0.599	0.001	<0.001	<0.001
Sex	0.075	0.203	0.510	0.295	0.297	0.984
Diagnostic Category						
Upper airway	0.593	0.824	0.061	<0.001	0.435	<0.001
Central nervous system	0.303	0.072	0.799	<0.001	0.233	<0.001
Musculoskeletal	0.436	0.591	0.574	0.001	0.005	0.236
Cardiopulmonary	0.635	0.225	0.056	0.080	0.397	<0.001
Other	0.598	0.957	0.531	0.999	0.868	0.551
Medical complexity						
Number of co-morbidities	0.863	0.083	0.847	<0.001	0.690	<0.001
Number of additional technologies	0.965	0.408	0.017	<0.001	0.531	<0.001
AT Surgery	0.256	0.145	<0.001	0.315	0.503	0.131

Any other major Surgery	0.330	0.470	<0.001	0.146	0.193	0.001
NIV Type	<0.001	<0.001	0.022	<0.01	0.300	<0.001
Site	0.269	<0.001	<0.001	0.257	0.027	0.083
Epoch	<0.001	0.018	0.030	0.014	<0.001	0.666
Trigger for NIV	0.202	0.243	<0.001	0.118	0.139	<0.001
Mask Type	0.517	0.466	0.005	0.053	0.044	0.005
Number of complications (first follow-up visit)	0.914	0.783	0.493	0.004 (+) correlation	0.043 (-) correlation	<0.001 (+) correlation
Number of complications (second follow-up visit)	0.148 (-) correlation	0.338	0.005 (+) correlation	0.018 (+) correlation	0.744	<0.001 (+) correlation
Underlying disease conditions						
CHD/Cardiac Failure	0.201	0.368	0.460	0.899	0.207	0.001
Congenital/Acquired Brain Injury	0.074	0.621	0.269	<0.001	0.996	<0.001
Chronic Respiratory Failure	0.308	0.380	0.030	<0.001	0.299	<0.001
SMA Type 1, 2 or 3	0.747	0.447	0.469	<0.001	0.466	0.009
Duchenne Muscular Dystrophy	0.681	0.076	0.792	0.001	0.255	0.216
Myopathy	0.212	0.040	0.374	0.122	0.261	0.209
Myotonic Dystrophies	0.198	0.970	0.015	0.530	0.907	0.315
Achondroplasia	0.040	0.454	0.341	0.212	0.007	0.433

Down Syndrome	0.179	0.216	0.554	0.150	0.750	0.212
Upper Airway abnormalities/PRS	0.738	0.057	0.400	0.158	0.146	0.860
Prader Willi Syndrome	0.415	0.523	0.751	0.687	0.160	0.618
Obesity/Diabetes/Metabolic Synd.	0.027	0.005	0.006	0.008	<0.001	0.029
Autism spectrum disorder	0.739	0.545	0.806	<0.001	0.216	<0.001
BPD	0.514	0.044	0.262	0.919	0.256	0.331

BPD – Bronchopulmonary dysplasia; CHD – Congenital heart disease; PRS – Pierre Robin Sequence; SMA – Spinal Muscular Atrophy

