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Respiratory functional and motor control deficits in children with spinal cord injury

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Abstract

Children with spinal cord injury (SCI) are at high risk for developing complications due to respiratory motor control deficits. However, underlying mechanisms of these abnormalities with respect to age development and injury characteristics are unclear. To evaluate the effect of SCI and age on respiratory motor control in children with SCI, we compared pulmonary function and respiratory motor control outcome measures in healthy typically developing (TD) children to age-matched children with chronic SCI. We hypothesized that the deficits in respiratory functional performance in children with SCI are due to the abnormal and age-dependent respiratory muscle activation patterns. Fourteen TD (age 7 ± 2 yrs., Mean $\pm$ SD) and twelve children with SCI (age 6 ± 1 yrs.) were evaluated by assessing Forced Vital Capacity (FVC); Forced Expiratory Volume in 1sec (FEV₁); and respiratory electromyographic activity during maximum inspiratory and maximum expiratory airway pressure measurements (PI_{max} and PE_{max}). The results indicate a significant reduction ($p < .01$) of FVC, FEV₁ and PE_{max} values in children with SCI compared to TD controls. During PE_{max} assessment, children with SCI produced significantly decreased ($p < .01$) activation of respiratory muscles below the neurological level of injury (rectus abdominus and external oblique muscles). In addition, children with SCI had significantly increased ($p < .05$) compensatory muscle activation above the level of injury (upper trapezius muscle). In the TD group, age, height and weight have significantly ($p < .05$) contributed towards increase in FVC and FEV₁. In children with SCI, only age was significantly ($p < .05$) correlated with FVC and FEV₁ values. These findings indicate the degree of SCI-induced respiratory functional and motor control deficits in children are age-dependent.

Keywords

Spinal cord injury; Respiratory function; Respiratory muscles; Pediatrics

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1. Introduction

In healthy individuals, spontaneous ventilation is driven by the respiratory centers located in the brain stem, modulated by respiratory muscle performance in response to the respiratory load (Feldman and Del Negro, 2006; Hess et al., 2013). However, in patients diagnosed with neuromuscular diseases, ventilation is compromised as respiratory muscles are unable to fully overcome the resistance associated with respiration (De Vivo et al., 1999). In adults with spinal cord injury (SCI), symptoms of respiratory insufficiency highly correlate with level and severity of the spinal lesion. Thus, cervical and thoracic injuries cause greater weakness or paralysis of respiratory muscles directly increasing the workload of breathing and leading to low spirometric volumes and static mouth pressures (Schilero et al., 2014; Schilero et al., 2009; Terson de Paleville and Lorenz, 2015). Consequently, impaired expiratory and abdominal muscle activation results in ineffective cough, mucus retention, and increased risk for atelectasis and pneumonia (Inal-Ince et al., 2009; Khirani et al., 2014; Schilero et al., 2009).

In typically developing (TD) children, respiratory muscle strength is a function of age (Wagener et al., 1984; Wilson et al., 1984). Neuromuscular diseases can hamper typical development of the trunk and respiratory muscles and can potentially lead to severe respiratory insufficiency (Chng et al., 2003; Dearolf et al., 1990; Fauroux and Khirani, 2014; Mulcahey et al., 2013; Wu et al., 2011): nearly all children who sustain SCI prior to skeletal maturity develop neuromuscular scoliosis, which decreases mechanical efficiency of the chest wall, further encumbering lung function (Lancourt et al., 1981; Mayfield et al., 1981; Mulcahey et al., 2013; Parent et al., 2010). As these children continue to develop and pathophysiological changes in the musculo-skeletal system progress, lung volumes and static mouth pressures continue to decline (Estenne et al., 1993; Fauroux and Khirani, 2014), because effects of SCI are exacerbated by the number of years of immobility and non-weight bearing (Schottler et al., 2012). Children with SCI are thus at high risk of developing and dying from respiratory complications; as such, it is crucial that evaluation and rehabilitation of respiratory function occurs as early as possible.

Pulmonary function testing with maximum inspiratory and expiratory pressure generation measurements are important tools used for diagnosis and monitoring of respiratory diseases (Jain et al., 2006; Stolzmann et al., 2008). However, these assessments fail to provide information about the underlying neural drive to the respiratory muscles. In adults, research has been conducted to evaluate respiratory muscle activation by using the respiratory motor control assessment (RMCA) (Ovechkin et al., 2010), a multi-muscle surface electromyographic (sEMG) approach to quantitatively evaluate respiratory muscle activation patterns in individuals with chronic SCI. There is a profound lack of knowledge about respiratory motor control development in TD children which creates a barrier to treating SCI-induced respiratory insufficiency in developing children. Therefore, the aim of this study was to evaluate respiratory motor function in TD children and in children with SCI by using standard pulmonary function testing in association with a respiratory multi-muscle sEMG recording. We hypothesized that deficits in respiratory functional performance in children with SCI are due to an abnormal and age-dependent respiratory muscle activation patterns.

2. Methods

2.1 Demographics and clinical characteristics

The Institutional Review Board (IRB) at the University of Louisville approved this study (IRB protocol #15.0585). Informed consent and assent were signed by legal guardians of children and by children above 7 years of age, respectively. Fourteen TD children (age 7 ± 2 years, 8 females and 6 males) with no history of respiratory or cardiovascular dysfunction, no thoracic-abdominal surgery, and no neuromuscular disease were recruited from the community and fourteen children with chronic SCI (age 6 ± 1 years, 6 females and 8 males) were recruited from Frazier Rehabilitation Institute, Louisville, KY. Human Locomotion Research Center Database in Louisville, KY (IRB approval # 06.0647) was used to recruit children with SCI. The severity of SCI in children was classified using the American Spinal Injury Association Impairment Scale (AIS) based on International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI) (for children above 6 years) and according to their medical records (for children below 6 years) (Mulcahey et al., 2011) (Table 1).

2.2 Pulmonary function testing (PFT)

Prior to testing, all children were measured for height and weight without shoes. In TD children, height and weight was measured in standing position whereas in children with SCI, recumbent board (supine) and electronic scale (sitting wheelchair scale) was used to measure height and weight, respectively. Standard spirometry measurements (Miller et al., 2005) were performed in the sitting position using Breeze suite System 2007 (Med-Graphics, St. Paul, MN) according to the ATS guidelines for children (Beydon et al., 2007). Values are reported as actual volume (Liters) and percentage of predicted for each child (Table 1). Tests occurred in a child-friendly environment by using instructions modified as needed (Beydon et al., 2007; Wagener et al., 1984). For FVC and FEV₁ measurements, children were instructed to place the mouthpiece in his or her mouth, breathe normally (tidal volume), then take a maximum deep breath in and blow out as fast and as long as possible. Computer incentive games (Med-Graphics, St. Paul, MN) were used during FVC and FEV₁ measurements: children were shown interactive birthday candles on a computer screen and were encouraged to continue until they “blow all the candles out” or until they could no longer blow. The flow-volume curves were analyzed by the examiner and were accepted or rejected based on ATS criteria for maximum expiratory maneuvers in children (Aurora et al., 2004; Beydon et al., 2007). Testing was repeated until three acceptable spirograms were obtained with one-minute rest between maneuvers; test duration was limited to a maximum of 15 minutes (Beydon et al., 2007). The highest values of FVC and FEV₁ were included for statistical analysis.

2.3 Maximum airway pressure measurements

Maximum inspiratory pressure (PI_{max}) and maximum expiratory pressures (PE_{max}) were measured by using a Differential Pressure Transducer (MP45-36-871-350), UPC 2100 PC card from Validyne Engineering (Northridge, CA). Both PI_{max} and PE_{max} were performed in sitting upright back-supported position with hip and knee joint flexed to 90 degrees and feet on ground. PI_{max} was recorded during maximum inspiratory effort at residual volume and

$PE_{\max}$ was recorded during maximum expiratory effort from total lung capacity using a three-way valve system with tube mouthpiece and 1.5mm diameter leak to prevent glottis closure and reduce the contribution from buccal muscles. Children were asked to maintain the pressure for at least two seconds; the highest mean value during 1 sec of each maneuver was used for statistical analysis.

2. 4 Surface electromyography (sEMG)

During $PI_{\max}$ and $PE_{\max}$ measurements, sEMG signals from respiratory muscles were recorded bilaterally using bipolar pre-amplified electrodes (Motion Lab Systems, Baton Rouge, LA). Skin area over the muscle belly was cleaned by alcohol swabs and electrodes were placed over the following muscles: upper trapezius (UT) at midclavicular line; pectoralis major (PEC) at midclavicular line; external intercostal (INT) at 6th intercostal space on axillary line; rectus abdominus (RA) at umbilical level; external oblique (OB) at midaxillary level; thoracic paraspinal (PST) at T₁₀ level; and lumbar paraspinal (PSL) at L₅ level. Ground electrodes were placed bilaterally on shin of tibia. Latex-free, hypoallergenic woven tape (BSN medical) was used to secure the electrodes in place. The sEMG signals were sampled at 2000Hz and mean rectified for burst duration and root mean square values were calculated (Aslan et al., 2013).

2.5 Statistical analysis

Data in table are presented as the mean $\pm$ (SD) and data in figures are represented in mean $\pm$ (SEM). Absolute values of FVC and FEV₁ in TD children and children with SCI were included for data analysis. A nonparametric, general linear mixed model was applied to test effects of injury on respiratory functional measures. Pearson correlation coefficient was used to examine a relationship between age, height, weight and pulmonary outcomes. Data were analyzed in SAS and plotted with R toolset (R Development Core Team, 2013).

3. Results

3.1 Pulmonary function test (PFT)

Children in both groups were matched (Table 1). No significant between group difference exist for age, height, and weight. Fourteen TD children (age 7 ± 2 years, 8 females and 6 males) and fourteen children with chronic SCI were recruited for this study, but only twelve children (age 6 ± 1 years, 5 females and 7 males) with chronic SCI performed acceptable spirometry curves.

Compared to children with SCI, children in the TD group produced significantly greater FVC and FEV₁ ($p < .01$) (Figure 1). In addition, TD children showed a strong, positive correlation between age and FVC ($r=.94$) and FEV₁ ($r=.82$) (Figure 2). Children with SCI also showed strong and positive correlations between age, FVC and FEV₁ ($r=.76$ and $r=.79$, respectively) (Figure 2). In TD children, a strong positive correlation was observed between FVC and height (0.97), and weight (0.78) and between FEV₁ and height (0.87), and weight (0.85). In the SCI group, height and weight did not significantly ($p > .05$) correlate to FVC or FEV₁ values.

3.2 Maximum airway pressure measurements

Children in the TD group generated significantly higher PE_{max} ($p < .01$) than children in the SCI group. No significant difference in PI_{max} was observed between the two groups (Figure 3). For children in both groups, age was not significantly correlated to PE_{max} values. In TD children, height ($r=0.68$) and weight ($r=0.35$) were significantly correlated with PE_{max} ($p < .05$); however, height and weight did not significantly correlate to PE_{max} in children with SCI.

In both groups, age was not significantly correlated to PI_{max} values ($p > 0.5$). In TD children, height ($r=0.39$) and weight ($r=0.17$) contributed significantly ($p < .05$) to the generation of PI_{max} . However, in children with SCI, height and weight were not significantly correlated with PI_{max} values.

3.3 Surface electromyography (sEMG)

During PE_{max} assessment, children with SCI produced significantly lower ($p < .05$) muscle activation of RA and OB and significantly higher ($p < .05$) activation for UT compared to children in the TD group (Figures 4 and 5). No significant differences ($p > .05$) in muscle activation of PEC, INT, PST, and PSL were found. During PI_{max} measurement, no significant differences ($p > .05$) were found between sEMG activation of respiratory muscles obtained from children in the TD and children in SCI groups (Figure 6).

4. Discussion

This is the first study undertaken to evaluate respiratory motor function in TD children and children with SCI by using a multi-muscle sEMG recording during PI_{max} and PE_{max} maneuvers to assess respiratory motor function (Ovechkin et al., 2010; Sherwood et al., 2000). Standard spirometric measurements, essential for clinical evaluations, are rarely used in preschool children (3–5 years of age) due to the notion that they are unable to perform a valid forced spirometer maneuver. However, evaluation of respiratory function in this age group is essential for clinical reasons and due to considerable growth and development of the respiratory system that occurs (Lanteri and Sly, 1993). In fact, only two children from this study who could not perform acceptable FVC and FEV_1 maneuvers were excluded from data analysis. Our study confirmed that valid spirometry curves can be obtained in preschool children, suggesting its feasibility in both younger and older children (Jeng et al., 2009; Nystad et al., 2002; Pesant et al., 2007; Zapletal and Chalupová, 2003).

In the present study, we found a significant reduction in FVC and FEV_1 values in children with SCI when compared to the age-matched TD controls. This reduction in FVC in children with SCI could be due to motor control deficits, increased stiffness of the chest wall due to the neuromuscular scoliosis (Birnkranz, 2002; Fauroux and Khirani, 2014; Dearolf et al., 1990; Mulcahey et al., 2013; Parent et al., 2010). Consistent with previous findings (Wagener et al., 1984), in the TD group, age, height, and weight each demonstrated a significant relationship with FVC; for FEV_1 , showing a significant positive correlation between respiratory functional measures and level of musculoskeletal development associated with age. However, in the SCI group, only age was significantly correlated with

FVC and FEV₁ values suggesting that SCI at any age would result in lower respiratory functional capacity.. Therefore, children who are injured at an early age are at higher risk of developing respiratory complications than those injured after they have attained skeletal maturity.

Children in the SCI group produced significantly lower PE_{max} values compared to children in the TD group due to the decreased activation of expiratory muscles like RA and OB innervated by thoracic and lumbar spinal segments (Holstege et al., 1987; Iizuka, 2011; Iscoe, 1998). Greater activation of PEC, INT, OB, and RA muscles was observed in TD children while children with SCI produced minimal or no activation of RA and OB muscles during PE_{max} maneuver. In TD children, intact innervation to respiratory muscles leads to adequate contraction of muscles against the respiratory load. However, SCI resulting in paresis or paralysis of these muscles, contributed to reduced lung volumes and expiratory airway pressure as when respiratory load exceeds the ability of the muscles (Fauroux and Khirani, 2014). In children with SCI, during PE_{max} assessment, activation of the UT (above the level of the lesion) was significantly greater than for children in the TD group, suggesting compensatory activation due to insufficient activation of primary muscles located below the level of the lesion. However, this adaptation was insufficient as it did not improve the PE_{max}. These results are consistent with studies that also reported compensatory strategies by adults with SCI to counteract the lack of neural activation of the primary muscles for expiration (Ovechkin et al., 2010; Terson de Paleville and Lorenz, 2015). Decreased PE_{max} values in the SCI group due to weak or paralyzed expiratory muscles are associated with a weak cough (McBain et al., 2013), leading to difficulty in clearing secretions from lungs, which causes mucus retention, atelectasis, and other significant respiratory complications (Schilero et al., 2009). Since the injury levels in our SCI group were below the level of phrenic nerve origination, there were no significant differences between the two groups for PI_{max} values and associated inspiratory sEMG activity measures (Zimmer et al., 2008). Children in both groups produced activation in UT, INT and PST muscles during PI_{max} maneuver.

The PE_{max} value in TD children was significantly correlated to height and weight, suggesting an increase in airway pressure with musculoskeletal growth, including respiratory muscles. This increase in the size of muscles is associated with a change in fiber composition, fiber size, and oxidative capacity and contraction properties of respiratory muscles, especially the diaphragm (Sieck G, 1991). However, in children with SCI, PE_{max} was significantly correlated with age, but not height and weight. SCI in children during the phase of growth and development results in an altered or atypical pattern of breathing due to weakness or paralysis of respiratory muscles and increased abdominal compliance. The combination of reduced lung compliance, increased chest wall stiffness and increased abdominal wall compliance lead to the reduced ventilation and increased work of breathing in association with respiratory muscles fatigue (De Troyer; Estenne; Estenne; Schilero et al., 2009).

In children with SCI, multi-muscle sEMG recording is essential to understand the involvement of different respiratory muscles. In addition, depending on level and severity of the SCI, the numbers of respiratory muscles involved may vary. Respiratory muscles are not

only involved during breathing, but also in maintaining posture control (Massery, 2005). Therefore, paralysis of respiratory muscles in children with SCI may also results in impaired posture. The ISNCSCI scale used in the clinic to assess level and severity of SCI does not include evaluation of these muscles (Mulcahey et al., 2011). Therefore, assessment of respiratory motor function using the RMCA protocol is useful in improving our knowledge and understanding of the underlying pathology.

Measurement of respiratory functions in children with neuromuscular disorders has been recommended by ATS and it also serves as a useful marker to assess the severity of disease and its prognosis over time (American Thoracic Society/European Respiratory, 2002). Our study indicates that not only can these functions be measured in TD preschool children, but also in children with SCI. This work provide foundation to assess the impact of therapeutic respiratory restorative interventions in children with SCI.

Conclusion

The purpose of this study was the comparative evaluation of respiratory motor function in children with SCI. We demonstrated that SCI-induced decrease in respiratory functional capacity in children is age-dependent and is associated with impaired respiratory muscle activation. Evaluation of respiratory function in conjunction with multi-muscle sEMG can be used as a tool to assess the impact of various rehabilitative interventions in children with SCI.

Study limitations

We did not control for rehabilitative interventions and medications that children with SCI were receiving which could have influenced their performance during testing. Studies in adults with SCI have suggested a positive relationship between level of injury and respiratory motor control outcomes, i.e., the higher the level of injury, the more significant reduction in pulmonary function parameters (Ovechkin et al., 2010; Schilero et al., 2009; Terson de Paleville and Lorenz, 2015). However, due to small sample size, we were not able to assess these relationships between levels of injury and respiratory motor control outcomes.

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Figure 1.

A. Forced Vital Capacity (FVC) and **B.** Forced Expiratory Volume in one second (FEV_1) in typically developing (TD) children and in children with spinal cord injury (SCI). *Note that FVC and FEV_1 (Mean $\pm$ SD) were significantly higher ($*p < .05$) in TD children compared to children with SCI.*

Figure 2.

Linear correlation between **A.** Forced Vital Capacity (FVC) and **B.** Forced Expiratory Volume in one second (FEV₁) and age in typically developing (TD) children and in children with spinal cord injury (SCI). *Note a strong linear relationship between FVC, FVC₁, and age in TD and SCI children.*

Figure 3.

A. Maximum expiratory pressure (PE_{max}) and **B.** Maximum inspiratory pressure (PI_{max}) in typically developing (TD) and spinal cord injury (SCI) groups. *Note that TD children produced significantly higher PE_{max} (Mean ± SD) when compared to children with SCI (*p=0.005).*

Figure 4.

A. Surface electromyographic (sEMG) activity of respiratory muscles and airway pressure during the Maximum Expiratory Pressure Task in normally developed (ND) individual and **B.** individual with SCI. Initiation of the task is marked by the dashed lines. *Note decreased airway pressure in SCI subject. Note also that activation of the expiratory muscles below level of injury is decreased [Right Intercostal (RINT)] and absent [Right Rectus Abdominus (RRA) and Right Oblique Abdominus (ROB)] with increased activity of the muscles above the level of SCI [Right Upper Trapezius and Right Pectoralis (RUT and RPEC)].*

Figure 5.

Surface electromyographic (sEMG) amplitude of respiratory muscles during maximum expiratory pressure (PE_{max}) measurements in typically developing (TD) and children with spinal cord injury (SCI). *Note that rectus abdominis (RA) and external oblique (OB) muscles activation was significantly higher in TD children compared to children with SCI (* $p=.04$ and * $p=.02$, respectively). Also note that children in SCI group produced significantly higher activation in upper trapezius (UT) (* $p=.01$) muscle.*

Figure 6.

Surface electromyographic (sEMG) amplitude of respiratory muscles during maximum inspiratory pressure (PI_{max}) measurements in typically developing (TD) and children with spinal cord injury (SCI). Note that no significant differences were observed for upper trapezius (UT) ($p=.051$), pectoralis major (PEC) ($p=.69$), intercostal (INT) ($p=.38$), thoracic paraspinal (PST) ($p=.86$), rectus abdominus (RA) ($p=.66$), external oblique (OB) ($p=.55$) and lumbar paraspinal (PSL) ($p=.28$) between TD and children with SCI.

Table 1

Demographics of Typically Developing Children (TD)

Subject (ID)	Age months	Gender	Height (cm)	Weight (kgs)	P _E max (cmH ₂ O)	P _I max (cmH ₂ O)	FVC (Liters)	FEV ₁ (Liters)	FVC% Predicted	FEV ₁ % Predicted
N149	41	F	89	17	19	-18	0.63	0.57	482	212
N150	57	M	114	20	42	-24	0.77	0.71	95	103
N133	48	M	98	17	33	NA	0.84	0.84	64	69
N130	61	F	114	20	53	-45	1.4	1.3	126	122
N134	60	M	106	17	66	-44	1.2	1.1	126	126
N145	83	F	114	27	63	-41	1.1	1.1	90	97
N126	77	M	101	17	44	-68	1.2	1.1	159	153
N110	94	F	122	27	26	-56	1.4	1.3	72	80
N127	97	F	124	23	49	-34	1.6	1.4	100	100
N147	119	F	144	37	58	-52	2.5	2.2	103	107
N146	122	F	137	51	42	-35	1.9	1.7	97	103
N144	129	F	129	33	65	-64	2	1.9	108	110
N108	138	M	145	27	70	-57	2.5	1.2	98	54
N109	146	M	160	44	72	-44	2.9	2.2	92	81
Mean and SD	90±33	8F,6M	120±19	26±10	51±14	-40±14	1.5±0.6	1.3±0.5	131±99	110±36

P_Emax=maximum expiratory pressures, P_Imax=maximum inspiratory pressures, FVC=forced vital capacity, FEV₁=forced expiratory volume in one second.

Table 2

Demographics of Children with Spinal Cord injury (SCI)

Subject (ID)	Age months	Gender	Height (cm)	Weight (kgs)	PEmax (cmH ₂ O)	PI _{max} (cmH ₂ O)	FVC (Liters)	FEV1 (Liters)	FVC% Predicted	FEV1% Predicted	Injury level	Time since injury (months)
P3	50	M	97	14	30	-28	0.59	0.59	103	109	T2	50
P7	72	F	114	20	13	-19	0.59	0.58	50	54	C5	30
P8	59	F	112	28	25	-29	0.75	0.68	78	66	T2	11
P9	71	F	114	23	30	-46	1	1	103	105	C5	58
P14	70	M	101	15	31	-41	0.8	0.71	103	98	T2	28
P16	65	M	109	19	58	-54	1.5	1.2	128	122	T12	17
P13	81	M	118	21	34	-18	1.4	1.3	100	101	T3	7
P4	76	F	101	14	28	-40	0.93	0.84	149	127	C8	73
P6	94	M	124	24	34	-46	1.7	1.5	106	109	T2	41
P1	118	M	148	31	65	-81	1.7	1.5	63	67	T1	69
P10	112	M	141	30	76	-69	1.4	1.2	59	61	C5	113
P5	128	F	137	32	48	-29	2	1.8	95	101	C5	129
Mean and SD	83±23	5F,7M	118±15	22±6	40±17	-42±18	1.2±0.4	1.1±0.4	94±27	93±23	NA	52±37

PE_{max}=maximum expiratory pressures, PI_{max}=maximum inspiratory pressures, FVC=forced vital capacity, FEV₁=forced expiratory volume in one second.