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Autonomic Dysfunction in Children with Cerebral Palsy, Static Encephalopathy, and Similar Conditions by Susan Agrawal

Many children with cerebral palsy, static encephalopathy, and similar conditions experience significant dysfunction of their Autonomic Nervous Systems (ANS). The ANS is the part of the nervous system that controls typically “automatic” functions of the body, such as temperature, heart rate, blood pressure, or digestion. Despite Autonomic Dysfunction being very common and severely affecting the quality of life of many children, there has been virtually no research in this area at all.

I did a simple search for the terms “cerebral palsy” and “dysautonomia,” a term used to describe poor function of the ANS, and came up with only six results. Five were published before 1990, and three of those were from the 1960s and 1970s. A search for “cerebral palsy” and the more general term “autonomic” still only yielded 36 articles in English, and only eight of these actually discussed autonomic symptoms. There were more results when searching for encephalopathy, but virtually none were relevant to static encephalopathy, cerebral palsy, or brain malformations. Most focused on infectious encephalopathies. A survey of medical textbooks on cerebral palsy and encephalopathy yielded little more than a few paragraphs on reflux, constipation, bladder issues, and temperature regulation problems.

Of the research that does exist, each article discusses only one parameter, such as heart rate, cold hands, or temperature regulation. There is no literature currently evaluating the prevalence of Autonomic Dysfunction or the correlation between severity of cerebral palsy/encephalopathy and Autonomic Dysfunction.

The Purpose and Structure of the Survey

In order to remedy this situation, we created a preliminary survey for children with cerebral palsy and similar conditions to evaluate the frequency of autonomic symptoms and likely Autonomic Dysfunction. We received a total of 85 parent-completed surveys, and 65 of these were ultimately included in the analysis. 20 surveys were disregarded, all due to a diagnosis other than cerebral palsy that is either progressive or is already linked to autonomic problems. The most common disregarded diagnoses were mitochondrial encephalopathy and Rett syndrome. Children who were diagnosed with cerebral palsy, static encephalopathy, hypoxic ischemic encephalopathy, anoxic brain injury, and any form of congenital brain malformation were included in the results.

Parents/guardians were initially questioned about the level of physical impairment their child experiences. The majority of responses came from children with severe physical impairment (Gross Motor Function Classification System [GMFCS] level V), with 43/65 or 66% of children in this category, as shown in Table 1. A much smaller number of responses was received for GMFCS categories I, II, III, and IV.

Table 1: Level of Physical Functioning

GMFCS	Description	#	%
I	Child walks without limitations; infant crawls and pulls to a stand	2	3%
II	Child walks with limitations; infant sits and creeps	10	15%
III	Child walks with a mobility device (walker, gait trainer); infant sits with support and rolls	6	9%
IV	Child self-propels wheelchair or uses a powerchair; infant has head control but not trunk control	4	6%
V	Child requires a manual wheelchair; infant lacks head or trunk control	43	66%

The remaining questions asked about specific autonomic symptoms grouped into categories in order to develop a total Autonomic Symptom Score for each child. Categories included temperature regulation, vital signs, GI symptoms, urological symptoms, and other symptoms. Some categories had point maximums to limit over-emphasis of a particular category. For example, while it was possible to earn more than 10 points for GI symptoms, no child was assigned a score over 10 points to prevent over-emphasis. The categories and points are shown in Table 2.

Table 2: Categories

Category	Number of Points
Temperature regulation	0 to 4 points
Vital signs	0 to 10 points
GI symptoms	0 to 10 points
Urological symptoms	0 to 5 points
Other symptoms	0 to 35 points
total	0 to 64 points

The subtotals from each category were then added up to create an Autonomic Symptom Score, which could range from 0 to 64. The specific items that were scored are shown in Table 3. Parents/guardians were also given the option of adding additional symptoms or descriptions that were scored on an individual basis.

Table 3: Autonomic Symptoms Scoring

Temperature regulation	0 to 4 points based on severity
Vital signs	0 to 2 points for each parameter, based on severity
	Blood pressure
	Heart rate
	Blood sugar
	Respirations
	Oxygen saturations
GI problems	A maximum of 10 total points
	1 point was earned for each of the following: reflux, mild/moderate constipation, delayed gastric emptying, bloating, other motility problem, or an NG/G tube.
	2 points were earned for each of the following: frequent retching/vomiting, severe constipation, dysmotility, feeding tube into the intestine, ACE/Malone, or colostomy.
	3 points were earned for each of the following: obstruction or an ileostomy.
	4 points were earned for TPN or IV fluids.
Bladder problems	A maximum of 5 total points
	1 point was earned for each of the following: neurogenic bladder, voiding dysfunction, retention, bladder spasms, kidney reflux, frequent infections, or other bladder problems.
	2 points were earned for each of the following: daily cathing or a cathing stoma.
	3 points were earned for each of the following: use of a Foley, suprapubic catheter, or vesicostomy.
Other problems	A maximum of 35 points
	2 points were earned for each of the following: dilated or pinpoint eyes, dry eyes, lack of sweating, profuse sweating, hormone/endocrine problems, inability to sense temperature on skin, fainting or dizziness, facial flushing, or circulation problems.
	3 points were earned for each of the following: problems with cortisol levels, neuropathic/visceral pain, or peripheral neuropathy.
	5 points were earned for autonomic crises or sympathetic storms.

It is important to note that each symptom alone may or may not be indicative of a problem with the Autonomic Nervous System. For example, while abnormal vital signs such as heart rate or blood pressure are often signs of Autonomic Dysfunction, they may also be abnormal in children with heart conditions or kidney problems. Similarly, common GI problems including reflux, vomiting, and constipation, may be caused by abnormal muscle tone and not dysfunction of the Autonomic Nervous System. But when a child exhibits multiple autonomic symptoms affecting multiple systems in the body, then Autonomic Dysfunction becomes a likely diagnosis.

Results of the Survey

Children who participated in the survey scored Autonomic Symptom Scores between 0 and 58. The average score was 17 and the median score was 13. Virtually all children in the survey had a positive Autonomic Symptom Score, though the majority fell in the mild range with few autonomic symptoms or mild Autonomic Dysfunction. Children who scored between 0-10 were not designated as having Autonomic Dysfunction because their symptoms were typically very mild. Anyone scoring 11 and above was designated as having Autonomic Dysfunction, including mild (11-20), moderate (21-30), moderately severe (31-40), and severe (41+).

21 of 65 (32%) children had only a few autonomic symptoms (1-10 points), 23 of 65 (35%) children fell in the mild range of autonomic dysfunction (11-20 points), 12 of 65 (18%) children were in the moderate range (21-30), 6 of 65 (9%) were in the moderately severe range (31-40), and 2 of 65 (3%) had severe Dysautonomia, with scores in the 50s. One child had no autonomic symptoms. These results are shown in Table 4.

Table 4: Severity of Autonomic Symptoms or Dysfunction

	Number	Percentage
No Autonomic Symptoms (0 pts.)	1	2%
Some Autonomic Symptoms (1-10 pts.)	21	32%
Mild Autonomic Dysfunction (11-20 pts.)	23	35%
Moderate Autonomic Dysfunction (21-30 pts.)	12	18%
Moderately Severe Autonomic Dysfunction (31-40 pts.)	6	9%
Severe Autonomic Dysfunction (40+ pts.)	2	3%

Results by Level of Physical Impairment

The Autonomic Symptom Scores were correlated with the level of physical impairment to try to determine if children who are more involved physically had more autonomic symptoms. Because so few surveys were received for children in categories I, II, III, and IV, it is difficult to determine with any accuracy this relationship. In addition, one child in Level II and another in Level V had exceptionally high Autonomic Symptom Scores of

58 and 52 respectively, perhaps due to secondary conditions, which makes it difficult to present an overall average. After excluding these two children, who skewed the results dramatically, the results shown in Table 5 were obtained. Children with mild impairment (levels I and II) are grouped together, as are children with moderate impairment (levels III and IV).

Table 5: Average Autonomic Symptom Scores by GMFCS Level (excludes two children with severe Dysautonomia)

GMFCS	Temp Reg	Vitals	GI	Uro	Other	TOTAL
I and II	1.3	2.5	3.9	0.8	4.6	13.2
III and IV	1.1	2.5	5.2	0.6	4.5	13.9
V	1.9	3.1	5.7	0.8	5.4	16.9

As can be seen in Table 5, there was a correlation between level of physical impairment and the likelihood of experiencing autonomic symptoms. Children with mild and moderate physical impairment (levels I, II, III, and IV) experienced very similar autonomic symptoms, with one exception, GI symptoms. Children with moderate impairment had significantly more GI symptoms than those with mild impairment. Children with severe impairment (level V), on the other hand, had slightly higher scores across all categories, leading to an overall higher score.

It is important to note that some autonomic symptoms, particularly GI symptoms, may also be influenced by muscle tone patterns. For example, the muscle tone of the GI sphincters and organs may be floppier, tighter, or fluctuating in children who have abnormal muscle tone. Therefore, the increase in GI symptoms seen in children with greater physical impairment may have more to do with increasingly abnormal muscle tone than Autonomic Dysfunction. For example, a child who has slightly low trunk tone but can walk is likely to have fewer GI problems than a child who has universally low muscle tone, including in the GI tract, and who requires a wheelchair.

While Table 5 looks at the average scores for each group, it is also interesting to examine individual children within each group to see if most children have mild or severe symptoms, as shown in Table 6. Children with mild physical impairment (GMFCS I and II) tended to just have a few mild autonomic symptoms (42%), but several children had moderate Autonomic Dysfunction (33%), and one had severe Dysautonomia. Children with moderate physical impairment (GMFCS III and IV) tended to also just have a few autonomic symptoms (50%), but some children had mild and moderate Autonomic Dysfunction (20% each), and one child had moderately severe Autonomic Dysfunction. Finally, children with severe physical impairment tended to have mild Autonomic Dysfunction (46%) or a few autonomic symptoms (26%), with a larger group of children in the moderately severe category (14%). Of the eight children with moderately severe or severe Autonomic Dysfunction, all but two (75%) were GMFCS V, with severe physical impairment.

Table 6: Level of Autonomic Symptoms or Dysfunction by Level of Physical Impairment

Mild Physical Impairment (GMFCS I and II)

	Number	Percentage
No Autonomic Symptoms (0)	1	8%
Some Autonomic Symptoms (1-10)	5	42%
Mild Autonomic Dysfunction (11-20)	1	8%
Moderate Autonomic Dysfunction (21-30)	4	33%
Moderately Severe Autonomic Dysfunction (31-40)	0	0%
Severe Autonomic Dysfunction (40+)	1	8%

Moderate Physical Impairment (GMFCS III and IV)

	Number	Percentage
No Autonomic Symptoms (0)	0	0%
Some Autonomic Symptoms (1-10)	5	50%
Mild Autonomic Dysfunction (11-20)	2	20%
Moderate Autonomic Dysfunction (21-30)	2	20%
Moderately Severe Autonomic Dysfunction (31-40)	1	10%
Severe Autonomic Dysfunction (40+)	0	0%

Severe Physical Impairment (GMFCS V)

	Number	Percentage
No Autonomic Symptoms (0)	0	0%
Some Autonomic Symptoms (1-10)	11	26%
Mild Autonomic Dysfunction (11-20)	20	46%
Moderate Autonomic Dysfunction (21-30)	6	14%
Moderately Severe Autonomic Dysfunction (31-40)	5	14%
Severe Autonomic Dysfunction (40+)	1	2%

Specific Autonomic Symptoms

Some autonomic symptoms were much more common than others. For example, 80% of children had some degree of difficulty regulating their body temperature, but only 26% ran a fever, became hypothermic, or had a life-threatening response to heat or cold. Similarly, 71% had trouble regulating at least one vital sign (blood pressure, heart rate, respirations, oxygen saturations, or blood sugar), but only 23% of these problems were severe. GI symptoms were almost universal, with 95% of children experiencing one or more symptoms, and 49% with moderate or severe symptoms. Urological symptoms were much less common, with 38% experiencing at least one, but only 17% with more than one symptom. Finally, the majority of children (75%) experienced at least one uncategorized autonomic symptom, but only 15% experienced a great number of additional symptoms. This information can be seen in Table 7.

Table 7: Specific Categories of Autonomic Symptoms Experienced

	Any Symptoms	Significant Symptoms
Temperature Regulation	52 (80%) [score of 1-4]	17 (26%) [score of 3 or 4]
Vital Signs	46 (71%) [score of 1-10]	15 (23%) [score of 6-10]
GI Issues	62 (95%) [score of 1-10]	32 (49%) [score of 6-10]
Urological Issues	25 (38%) [score of 1-5]	11 (17%) [score of 2-5]
Other Issues	49 (75%) [score >1]	10 (15%) [score of 11+]

Temperature regulation is a particularly important measure of Autonomic Dysfunction because there are few other causes for the inability to regulate one's temperature. The high number of children affected by this problem to some degree, 80%, is suggestive of how widespread Autonomic Dysfunction is in children with cerebral palsy and similar conditions. 26% of children were severely affected, and the likelihood of significant Autonomic Dysfunction is particularly high in these children.

When it comes to vital signs and measurements of health, the most common abnormalities, affecting more than half of children, were found in breathing rate (64%), heart rate (60%), and oxygen saturations (56%). Abnormalities in blood sugar were less common (26%), while abnormalities in blood pressure were relatively common (46%). Children with extremely abnormal vital signs were most likely to have an abnormal heart rate (31%) or breathing rate (26%). These results are shown in Table 8. Because many parents/guardians did not know their child's blood sugar, blood pressure, or oxygen saturations, the percentages in Table 8 only include children with a known result.

Table 8: Abnormalities in Vital Signs

	Abnormal	Very Abnormal
Blood Pressure	25 (46%) [score of 1-2]	8 (15%) [score of 2]
Heart Rate	37 (60%) [score of 1-2]	19 (31%) [score of 2]
Blood Sugar	12 (26%) [score of 1-2]	4 (9%) [score of 2]
Breathing Rate	39 (64%) [score of 1-2]	16 (26%) [score of 2]
Oxygen Saturations	33 (56%) [score of 1-2]	9 (15%) [score of 2]

GI symptoms were very common, with 82% of children experiencing reflux, 74% with constipation, 71% with an NG or G tube, and 57% with delayed emptying of the stomach. Symptoms that are common in severe Dysautonomia, such as dysmotility (15%), pseudo-obstruction (11%), and dependence on IV nutrition (9%) or a GJ/J/NJ tube (20%), were less common overall, and typically only seen in children with significant Autonomic Dysfunction. These results may be seen in Table 9. It is important to remember that many of these conditions may have alternate causes, such as muscle tone problems, and only suggest Autonomic Dysfunction in combination with other symptoms.

Table 9: GI Symptoms

	Number	Percentage
Reflux	53	82%
Constipation	48	74%
NG/G Tube	46	71%
Delayed Gastric Emptying	37	57%
Retching/Vomiting	31	48%
Bloating	20	31%
GJ/J/NJ Tube	13	20%
Dysmotility	10	15%
Pseudo-Obstruction	7	11%
TPN or IV Fluids	6	9%

Urological Symptoms were rather uncommon overall, and the majority of children affected had urine retention as their primary or only symptom (26%). More severe symptoms, such as voiding dysfunction (9%), bladder spasms (9%), and requiring catheterization (8%) were very uncommon. These results may be seen in Table 10.

Table 10: Urological Symptoms

	Number	Percentage
Urine Retention	17	26%
Neurogenic Bladder	8	12%
Frequent Infections	7	11%
Voiding Dysfunction	6	9%
Bladder Spasms	6	9%
Cathing	5	8%
Kidney Reflux	4	6%

The list of other unrelated autonomic symptoms is long, and most children did experience one or more symptoms. The most common were circulation problems (43%), facial flushing (39%), sweating issues (33%), and the inability to sense pain on the skin (25%). Less common symptoms, especially those typical of severe Dysautonomia, occurred rather infrequently, with 14% of children experiencing autonomic crises, 6% with peripheral neuropathy, and 3% with high or low levels of cortisol. These results may be seen in Table 11. While some of these symptoms are particularly suggestive of Autonomic Dysfunction, especially sweating issues, inability to sense pain or temperature, dilated/pinpoint eyes, and autonomic crises, other symptoms may have other causes. Circulation problems, for example, may be caused by high muscle tone and immobility in some children with cerebral palsy or similar conditions.

Table 11: Other Autonomic Symptoms

	Number	Percentage
Circulation Problems	28	43%
Facial Flushing	25	39%
Sweating Issues (profuse or not enough)	22	33%
Does Not Feel External Pain	16	25%
Dilated/Pinpoint Eyes	13	20%
Neuropathic/Visceral Pain	11	17%
Endocrine Issues	9	14%
Autonomic Crises	9	14%
Does Not Sense External Temperature	7	11%
Fainting/Dizziness	6	9%
Dry Eyes	4	6%
Peripheral Neuropathy	4	6%
Cortisol Issues	2	3%

Conclusion

These results suggest that Autonomic Dysfunction is very common in children with cerebral palsy and similar conditions, but is currently unrecognized and untreated in many children, especially those with the most severe physical impairment. Many children with moderate, moderately severe, or severe Autonomic Dysfunction have never been diagnosed with Dysautonomia or Autonomic Dysfunction. Only 3 out of 12 children with moderate Autonomic Dysfunction (25%) have been given this diagnosis. Children with moderately severe Autonomic Dysfunction were more likely to be given this diagnosis (4 out of 6 children, or 67%), and all children with severe Autonomic Dysfunction had been diagnosed.

This preliminary study does have many limitations. Most significantly, it relies on parent report, and some parents may have vastly different definitions of what constitutes severe symptoms. In addition, parents/guardians filled out the survey knowing what topic would be addressed, and it is possible that those whose children experience autonomic issues were more likely to respond. Finally, there was neither a control group nor a balanced number of children in each category of physical impairment, potentially skewing the results further. Nonetheless, this study suggests that much more research needs to be done in this area.

Because these symptoms can affect quality of life so dramatically, and some can be treated with great results, it is very important to recognize just how common Autonomic Dysfunction is in children with cerebral palsy and similar conditions. Hopefully, medical researchers will devote some time studying this relationship and finding ways to treat Autonomic Dysfunction.