



# American Academy of Developmental Medicine and Dentistry

## Developmental Medicine and Dentistry Reviews & Reports



**Message from the Editor:** A neurodevelopmental disorder (NDD) is defined as a precise genetic or acquired biological brain disorder or condition that is responsible for childhood-onset brain dysfunction. If a neurodevelopmental disorder is severe enough, it may cause a “developmental disability” e.g. mental retardation, cerebral palsy, epilepsy, or autism. When caring for patients with neurodevelopmental disorders (NDDs), it is important to consider the four primary categories of clinical manifestations or “complications” of these disorders (cognitive dysfunction, behavioral problems, motor dysfunction, and seizures) as part of a larger medical issue. Secondary to these “complications,” patients may also experience various “consequences” that will adversely affect their health.

A retrospective review was performed on 43 patient charts at Hunterdon Developmental Center in order to determine the various causes, complications, and consequences of childhood-onset brain dysfunction (COBD) in this population. The origins of NDDs are varied in this study

with 42 percent having unknown causes, 23 percent having genetic causes, and 35 percent having acquired causes. Also, patients in this study were found to have an average of 3.3 “complications” of NDD. With regards to secondary “consequences,” pneumonia, GERD, scoliosis, polypharmacy, dental concerns, and visual deficits affected over 50 percent of the sample population.

The message is clear; a patient’s childhood-onset brain dysfunction always has an underlying medical cause (even though a precise neurodevelopmental diagnosis may not have yet been determined). Physicians who practice in institutions and the community need to ascertain an accurate neurodevelopmental diagnosis and effectively evaluate and manage the various complications and consequences that result from that underlying neurodevelopmental disorder.

—Philip May, M.D., Co-Editor, R&R

# Causes, Complications, and Consequences of Neurodevelopmental Disorders Encountered in a Cohort of Adults with Childhood-onset Brain Dysfunction Who are Institutionalized

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### Introduction:

In their 1997 article, Carl Tyler and Claire Bourguet discussed the national trend to deinstitutionalize adults with intellectual disabilities and place them in community residential settings. As a result, community based primary care physicians (PCPs) would assume care for this new patient population. In their research, the authors gathered information on 21 adults at the time of deinstitutionalization and subsequently obtained follow-up data over the next three years from their new community based PCP.

At deinstitutionalization, each patient had an

average of 2.0 diagnoses related to chronic conditions with an average of 2.1 long-term medications. After one year of community care, they had an average of 2.9 diagnoses with an average of 1.7 long-term medications. Fifty percent of these diagnoses (which included hepatitis B, dysphagia, acid peptic disease, GERD, absence seizures, primary degenerative dementia, bronchiectasis, and idiopathic iridocyclitis) were newly formulated by the PCP. In addition, some patients who had chronic conditions while institutionalized no longer presented with them in community-based practice. For

example, one-third of patients previously diagnosed with chronic constipation did not present with this in the community. Similar results were seen with glaucoma, schizophrenia, dextrocardia, and cerebral palsy.

After one year of community-based practice, none of the patients had to be re-admitted to institutions. Although there were no quantitative comparisons made, many patients showed functional improvement in activities of daily living (most notably a patient who was wheelchair bound while institutionalized gained the ability

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to ambulate and previously incontinent patients gained urinary continence). The average weight of these patients increased by eight pounds with some gaining as much as 34 pounds. Other issues in care noted by the new PCPs included lack of immunization records in 38 percent of the patients, lack of cholesterol testing in 24 percent, and adverse health effects due to the use of cigarettes as rewards while institutionalized.

Based on their findings, the authors discuss six main recommendations that should be heeded in order to better care for this patient population, whether institutionalized or not. First, it is important for physicians to attempt to identify the cause of a patient's mental retardation so that one can be vigilant for syndrome-specific health complications that may occur. Second, look for conditions commonly seen in persons with mental retardation, such as hepatitis B, dysphagia, aspiration, and seizures. Third, think of geriatric syndromes (incontinence, falls, immobility, cognitive decline, osteoporosis) that may emerge at an earlier age in this population. Fourth, identify and treat secondary disabilities or consequences that may occur as a result of a patient's primary complications. Fifth, apply medication reduction strategies. Lastly, preventative health care strategies are essential and should be implemented in this population.

In the years following Tyler and Bourget's work, other studies have been published discussing the issues faced by patients with neurodevelopmental disorders. Baxter, et al. performed a similar study in Wales in which patients with "intellectual disabilities" (i.e. the cognitive complication of a neurodevelopmental disorder) were encouraged to see a primary care practitioner. Their data showed that of those patients that followed up, more than half had new health concerns diagnosed (63 percent had one concern, 25 percent had two, and

12 percent more than two). Henderson, et al., performed a study of patients with Down syndrome and reported that certain medical conditions occur more frequently in this group of adults. However, they discovered that follow-up of common complications that occur in individuals with Down syndrome is infrequent. About half of adults with Down syndrome had not seen a doctor in the previous 12 months and one-third had not visited a doctor in the previous three years.

It was with this work in mind that the following research project was begun. In order to be able to effectively care for patients with neurodevelopmental disorders, Tyler and Bourget's six recommendations should be considered for each individual. In the following research, a small cohort of patients at Hunterdon Developmental Center was followed and, with the above framework in mind, screened for different medical complications and consequences of their neurodevelopmental disability.

Patients with NDDs are a very heterogeneous group with various causes, complications, and consequences of their NDD. With regards to causes, there are three main categories: unknown, genetic, and acquired. As seen in Table 3, genetic causes include metabolic syndromes (e.g. PKU), structural disorders (e.g. tuberous sclerosis), and chromosomal abnormalities (e.g. Down syndrome). Acquired causes can be subdivided into prenatal (e.g. congenital rubella syndrome), perinatal (e.g. traumatic brain injury of birth), and postnatal disorders (e.g. lead encephalopathy). Regardless of the cause of NDD, these patients may display various combinations of four main complications: cognitive dysfunction (e.g. mental retardation), motor dysfunction (e.g. spasticity), behavioral issues (e.g. impulsivity), and seizures. Lastly, these four "complications" can result in numerous "consequences." For example, GERD is a consequence that can result from scoliosis,

which itself is a consequence of a motor complication. Therefore, the purpose of this study is to show that NDD patients do not simply have an isolated condition, but rather a broad spectrum of complications and consequences that are related to the initial cause of dysfunction (i.e. the neurodevelopmental disorder).

## Methods:

A retrospective chart review was completed on 43 patients currently residing in two inpatient units (referred to in this paper as Unit 1 and Unit 2) at the Hunterdon Developmental Center located in Clinton, NJ. The Hunterdon Developmental Center is a 650-bed residential facility for adults with neurodevelopmental disorders, complicated by cognitive dysfunction (mental retardation). In addition to cognitive dysfunction, roughly 30-40 percent of the residents of HDC receive psychotropic medication for a variety of severe destructive behaviors, 30 percent demonstrate complex seizures, and 30 percent demonstrate neuromotor dysfunction as well. A precise and accurate neurodevelopmental diagnosis has been established for approximately 30-40 percent. Established diagnoses include Down syndrome, Fragile X syndrome, Trisomy 8 mosaic syndrome, Trisomy 13 syndrome, William Syndrome, Velo-cardio-facial syndrome, Rubenstein-Taybi syndrome, Rett syndrome, Angelman syndrome, Smith Magenis syndrome, PKU, tuberous sclerosis, neurofibromatosis, Sturge Weber syndrome, congenital rubella syndrome, the syndrome of hypoxic/ischemic perinatal brain injury, lead encephalopathy, hydrocephalus, prosencephaly, schizencephaly, and other diagnoses. The Center is operated by the NJ Division of Developmental Disabilities, under the auspices of the New Jersey Department of Human Services, with funding provided through state appropriation and federal programs (ICF/MR).

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## INSTRUCTIONS

For CME Credit read the editorial above and the article below and complete the Content Test and CME Evaluation Form at the end. Please read "Information and Instructions" following the article.

Specific learning objectives for this CME activity (please refer to general objectives).

Upon completion of reading of this article the learner will be able to:

1. Define the term "neurodevelopmental disorder"
2. Define the term "complication" of a neurodevelopmental disorder

3. Define the term "consequence" of a complication of a neurodevelopmental disorder

Upon receipt and acceptance of the completed evaluation form/post-test, the AADMD CME Program will maintain on file a record for six years designating your credits earned. If you should need a written verification contact, Philip May, M.D. at 908-510-3062.

# Table 1: Patient Diagnoses and Complications

Patient #	Age/Gender	Diagnosis	Cognitive	Motor	Seizure	Behavior	Total Complications
<b>Unit 1</b>							
1	60 M	Down Syndrome	P			P	2
2	46 M	Arrested Hydrocephalus	P	P		P	3
3	64 M	Peripartum Ischemic Encephalopathy	P	P		P	3
4	71 M	Down Syndrome	P	P	P		3
5	36 M	Skull Fracture with Subdural Hematoma	P	P	P		3
6	44 M	Angelman Syndrome	P	P	P		3
7	70 M	Prematurity	P	P			2
8	60 M	Coffin-Lowry Syndrome	P	P	P	P	4
9	66 M	Unknown	P	P	P	P	4
10	58 M	Down Syndrome	P		P	P	3
11	72 M	Unknown	P	P	P		3
12	47 M	FG Syndrome	P	P	P	P	4
13	35 M	Unknown	P	P	P		3
14	26 M	Unknown	P	P	P	P	4
15	54 M	Unknown	P	P	P	P	4
Unit1 Totals:			15	13	11	9	3.2
Unit1 Percentages:			100%	87%	73%	60%	
<b>Unit 2</b>							
1	52 M	Prematurity	P			P	2
2	44 M	Anoxemia at Birth	P	P	P		3
3	51 M	Cerebral Migration Defect	P	P	P	P	4
4	43 M	Unknown	P	P	P	P	4
5	50 M	Smith-Magenis Syndrome	P	P	P	P	4
6	44 M	Unknown	P	P	P		3
7	39 M	Angelman Syndrome	P	P	P		3
8	43 M	Unknown	P	P	P		3
9	40 M	Unknown	P	P	P	P	4
10	45 M	Postnatal Viral Encephalitis	P	P	P		3
11	74 M	Prematurity	P	P	P	P	4
12	40 M	Unknown	P	P	P		3
13	41 M	Unknown	P	P	P		3
14	44 M	Postnatal Meningitis	P	P	P		3
15	35 M	Cytomegalic Inclusion Disease	P	P	P		3
16	45 M	Primary Cranial Defect	P	P		P	3
17	45 M	Prematurity	P	P	P	P	4
18	44 M	Unknown	P	P	P		3
19	37 M	Down Syndrome	P	P	P	P	4
20	43 M	Unknown	P	P	P		3
21	46 M	Unknown	P	P	P	P	4
22	36 M	Unknown	P	P	P		3
23	54 M	Tuberous Sclerosis	P	P	P		3
24	59 M	Primary Intracranial Neoplasm	P	P	P	P	4
25	75 M	Mechanical Injury at Birth	P		P	P	3
26	44 M	Postnatal Anoxemia	P	P	P	P	4
27	27 M	Fetal Distress Syndrome	P	P	P		3
28	43 M	Unknown	P	P			2
Unit 2 Totals:			28	26	25	13	3.3
Unit 2 Percentages:			100%	93%	89%	46%	
Overall Totals:			43	39	36	22	3.3
Overall Percentages:			100%	91%	84%	51%	

## Table 2: Consequences of Neurodevelopmental Dysfunction

### Unit 1

Patient Number	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	Total #	Total %	1	2	3	4	5	6	7
Pneumonia	✓	✓	✓	✓	✓	✓	✓		✓	✓	✓	✓	✓	✓	✓	14	93%	✓	✓	✓	✓	✓		✓
Chronic Lung Disease	✓					✓	✓				✓		✓	✓	✓	7	47%					✓		
Oropharyngeal Dysfunction	✓	✓	✓	✓	✓	✓			✓	✓		✓	✓	✓		11	73%			✓				
Feeding Tube	✓		✓		✓	✓	✓					✓	✓	✓		8	53%			✓				
GERD		✓	✓	✓	✓	✓	✓			✓	✓		✓		✓	10	67%	✓	✓		✓			
Barrett's Esophagus			✓			✓	✓								✓	4	27%		✓		✓			
Constipation	✓			✓	✓	✓	✓	✓			✓	✓	✓	✓	✓	11	73%				✓		✓	
Scoliosis					✓	✓	✓	✓	✓	✓	✓					7	47%		✓		✓	✓	✓	
Osteoporosis			✓	✓	✓		✓	✓		✓					✓	7	47%		✓		✓			
Fractures			✓	✓	✓	✓	✓	✓	✓		✓		✓			9	60%	✓		✓		✓	✓	✓
Polypharmacy	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓		✓	✓	✓	14	93%	✓		✓	✓	✓	✓	✓
Decubiti		✓									✓					2	13%							
Bladder Dysfunction											✓					1	7%	✓						
UTI	✓	✓	✓						✓		✓	✓				6	40%	✓		✓				
Dental Problems				✓			✓				✓					3	20%	✓	✓	✓	✓	✓	✓	✓
Visual Deficits		✓	✓	✓	✓		✓	✓		✓	✓	✓	✓		✓	11	73%		✓	✓	✓	✓	✓	✓
Hearing Deficits					✓											1	7%	✓						
Hepatitis B Carrier	✓	✓														2	13%							
Hypothyroidism			✓	✓						✓						3	20%				✓			
Hypertension							✓		✓		✓					3	20%	✓						
Hyperlipidemia				✓			✓		✓		✓					4	27%							

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Hunterdon Developmental Center employs a large staff, including nine full time physicians. A 70-bed infirmary is located in the center of the campus. The Center also provides skilled nursing, staff EMT's, 24-hour care and medical services, and a comprehensive rehabilitation program for physical therapy, speech therapy, audiology, and occupational therapy. In addition to routine medical services provided by staff physicians, board certified specialists offer regular clinics in psychiatry, neurology, epileptology, dermatology, gynecology, ENT, orthopedics, ophthalmology, and genetics. Collaborations with Warren Community Hospital and Hunterdon Medical Center provide for acute Hospital care when needed. The medical staff of Hunterdon Developmental Center also provides for regular teaching activities in developmental

medicine for residents, fellows, and medical students affiliated with the Robert Wood Johnson Medical School.

Information obtained from each patient's chart included a diagnosis (if available), occurrence of the four main complications of NDD (i.e. cognitive, motor, seizure, behavior), and occurrence of certain common health consequences. This data was then statistically analyzed to observe the prevalence of specific etiologic neurodevelopmental diagnoses and the complications and consequences of those NDDs in this population.

### Results:

The population being studied in this report consisted of 43 males whose ages varied from 27-75 years with a mean of 49. Table 1 shows the prevalence of the main "complications" of

NDD in this population. Overall, 100 percent of patients had a cognitive dysfunction, 91 percent had a motor dysfunction, 84 percent had seizures, and 51 percent had behavioral issues. Unit 1 patients had an average of 3.2 complications where Unit 2 patients had an average of 3.3. Motor dysfunction and seizures were more prevalent in Unit 2, whereas, behavioral issues were more common in Unit 1. Table 2 shows the prevalence of certain common "consequences". Overall, pneumonia, GERD, scoliosis, polypharmacy, dental problems, and visual deficits were seen in over 50 percent of the population. When examining each consequence individually, certain trends were observed. Pneumonia was evident in 93 percent of the patients in Unit 1 but was only seen in 57 percent of Unit 2 patients. Similarly, chronic lung disease (47 percent vs. 18 per-

## Unit 2

## Overall

	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	Total #	Total %	Total #	Total %
	✓	✓	✓		✓		✓			✓	✓	✓		✓							✓	16	57%	30	70%
				✓		✓			✓									✓				5	18%	12	28%
	✓														✓							3	11%	14	33%
																						1	4%	9	21%
	✓	✓	✓	✓	✓	✓			✓		✓		✓		✓	✓	✓					15	54%	25	58%
			✓	✓							✓		✓				✓					7	25%	11	26%
	✓	✓	✓								✓					✓						7	25%	18	42%
	✓	✓	✓	✓	✓	✓	✓	✓		✓		✓			✓	✓		✓		✓	✓	19	68%	26	60%
									✓	✓	✓		✓	✓							✓	8	29%	15	35%
					✓				✓	✓			✓	✓		✓				✓	✓	12	43%	21	49%
	✓	✓	✓	✓	✓	✓	✓	✓	✓		✓	✓	✓	✓	✓	✓		✓	✓	✓		24	86%	38	88%
										✓	✓							✓				3	11%	5	12%
					✓												✓					3	11%	4	9%
		✓			✓												✓	✓				6	21%	12	28%
	✓	✓	✓	✓		✓				✓	✓		✓	✓		✓	✓			✓		19	68%	22	51%
	✓	✓	✓	✓	✓	✓	✓	✓	✓			✓								✓		17	61%	28	65%
																		✓	✓			3	11%	4	9%
					✓													✓				2	7%	4	9%
									✓			✓						✓				4	14%	7	16%
																		✓				2	7%	5	12%
				✓								✓										2	7%	6	14%

cent), oropharyngeal dysfunction (73 percent vs. 11 percent), feeding tube placement (53 percent vs. 4 percent), constipation (73 percent vs. 25 percent), and urinary tract infections (40 percent vs. 21 percent) had prevalence rates that were much higher in Unit 1. Scoliosis (47 percent vs. 68 percent) and dental problems (20 percent vs. 68 percent) were the only two consequences seen at much higher rates in Unit 2. Table 3 shows the prevalence of various causes of NDDs. As a group, genetic disorders were the least common known causes (23 percent of cases), with Down syndrome (9 percent) being the most common single diagnosis within this set. Acquired disorders caused 35 percent of cases, with prematurity being the single most common cause (9 percent) of that group. Unknown causes comprised 42 percent of cases, and was therefore the most common cat-

egory. Thus, a precise etiologic diagnosis (i.e. a specific neurodevelopmental disorder) has not been established for most of our patients with a childhood-onset brain dysfunction.

### Discussion:

After reviewing the charts of these 43 patients, it is clear how NDDs present as a constellation of complications and consequences. Hunterdon Developmental Center provided for a sample size with a variety of NDD causes. The specific etiologic origins of NDDs were not evenly divided with 42 percent having unknown causes, 23 percent having genetic causes, and 35 percent having acquired causes. Also, patients in this study were found to average 3.3 complications of NDDs. With regards to secondary consequences, pneumonia, GERD, scoliosis,

polypharmacy, dental concerns, and visual deficits were the most common ones in this population.

The differences in the complications and consequences observed in Unit 1 versus Unit 2 can be explained by the manner in which Hunterdon divides their patients. For the most part, patients at this facility are assigned to a unit based on their functional abilities. Unit 1 is located in the Health Services Residence building and consists of patients with severe complications and medical consequences. This can be noted by the fact that this unit has a high incidence of pneumonia, chronic lung disease, oropharyngeal dysphagia, and feeding tube placement. In contrast, Unit 2 consists of patients who are non-ambulatory and more prone to seizures. Therefore, these

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### Table 3: Causes of Neurodevelopmental Dysfunction

Causes of Brain Dysfunction	Unit 1		Unit 2		Overall	
	Number	Percentage	Number	Percentage	Number	Percentage
<b>Unknown Cause</b>	<b>5</b>	<b>33%</b>	<b>13</b>	<b>46%</b>	<b>18</b>	<b>42%</b>
<b>Genetic Disorders</b>	<b>6</b>	<b>40%</b>	<b>4</b>	<b>14%</b>	<b>10</b>	<b>23%</b>
- Metabolic Disorders	0	0%	0	0%	0	0%
- Structural Malformations	0	0%	1	4%	1	2%
<i>Tuberous Sclerosis</i>	0	0%	1	4%	1	2%
- Chromosomal Abnormalities	6	40%	3	11%	9	21%
<i>Down Syndrome</i>	3	20%	1	4%	4	9%
<i>Angelman Syndrome</i>	1	7%	1	4%	2	5%
<i>Coffin-Lowry Syndrome</i>	1	7%	0	0%	1	2%
<i>FG Syndrome</i>	1	7%	0	0%	1	2%
<i>Smith-Magenis Syndrome</i>	0	0%	1	4%	1	2%
<b>Acquired Disorders</b>	<b>4</b>	<b>27%</b>	<b>11</b>	<b>39%</b>	<b>15</b>	<b>35%</b>
- Prenatal Disorders	1	7%	1	4%	2	5%
<i>Arrested Hydrocephalus</i>	1	7%	0	0%	1	2%
<i>Prenatal Infection</i>	0	0%	1	4%	1	2%
- Perinatal Disorders	2	13%	4	14%	6	14%
<i>Prematurity</i>	1	7%	3	11%	4	9%
<i>Peripartum Ischemic Encephalopathy</i>	1	7%	0	0%	1	2%
<i>Fetal Distress Syndrome</i>	0	0%	1	4%	1	2%
- Postnatal Disorders	1	7%	6	21%	7	16%
<i>Anoxemia at Birth</i>	0	0%	2	7%	2	5%
<i>Postnatal Infection</i>	0	0%	2	7%	2	5%
<i>Mechanical Injury at Birth</i>	1	7%	1	4%	2	5%
<i>Primary Neoplasm</i>	0	0%	1	4%	1	2%
<b>TOTAL</b>	<b>15</b>		<b>28</b>		<b>43</b>	

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patients had a higher number of motor and seizure complications but fewer consequences that required immediate medical attention.

The primary limitation of this study was its small sample size (n=43). Also, the charts reviewed were those of patients who were more medically ill (Unit 1) or more likely to be non-ambulatory (Unit 2) than a typical NDD patient, which introduced selection bias into this study. In addition, this was a retrospective study so it was subject to the bias of the original charting physician and staff. For future studies, it would be advised to perform a cohort study consisting of the entire Hunterdon Developmental Center population. This would give a better indication of the

prevalence of NDD causes, complications, and consequences in the average patient.

Despite the study's limitations, its underlying message should be clear: a patient's childhood-onset brain dysfunction always has an

underlying medical cause that should be sought. With this mindset, members of the healthcare community would be better able to anticipate and treat the resulting complications and consequences. •

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