Supplemental Information

DETAILS ON STUDY DATA SOURCES AND COVARIATES

In Ontario, all interactions with the health care system are captured in health administrative data sources that are housed at ICES. Encoded identifiers allow linkage of multiple data sources at ICES so individuals can be managed over time. Data sources used in this study included the Discharge Abstract Database for hospitalization records, same-day surgery data for outpatient procedures, the Registered Person Database and Ontario Census Area Profiles for demographic data, Ontario Registrar General Database for vital statistics, Ontario Health Insurance Plan billing for outpatient and emergency department visits, the Ontario Mother-Baby linked data set and the Citizenship and Immigration Canada database for immigration status, National Ambulatory Care Reporting System for emergency department visits, and the ICES Physician Database for physician specialty. All data sources use deterministic linkage except the Ontario Registrar General, which requires probabilistic linkage (96.2% linkage rate). The study protocol and programming code are available from the corresponding author (K.E.N.).

We identified demographic and clinical characteristics that might impact rates of health care use or be associated with more severe NI using the framework for health care use created by Aday and Andersen.[26]

Tube Type

GJTs are often preferentially used for children with reflux disease causing morbidity, so we differentiated between enteral feeding tube types. Feeding tubes were classified as GTs or GJTs on the basis of the physician billing codes (occurring within 7 days of the hospital procedure code to account for common lag time in physician billing) or the procedure code when the billing code was ambiguous for tube type.

Clinical Characteristics at Time of Procedure

Children who require feeding tube placement earlier in life may have more severe NI than older children and be at higher risk of hospitalization. Being an inpatient at the time of feeding tube placement may be another marker of medical fragility. Similarly, children who have severe enough reflux disease to warrant an antireflux procedure (GJT or antireflux surgery) may be more severely affected.

Type of NI

NI was determined by using the diagnosis code(s) present on the hospitalization record closest to the time of the feeding tube placement. We used the subcategories from Feudtner's well-established list of CCCs for neurologic diagnoses (eg, brain and spinal cord malformations) and the categories from that list for nonneurologic diagnoses associated with NI (eg, metabolic diseases).[21] However, given the possible range of severity and trajectories within a specific category (eg, cerebral palsy), we did not stratify outcomes on the basis of subcategories or categories. Instead, 3 general pediatricians (K.E.N., A.G., and E.C.) determined whether each NI diagnosis code was considered static or progressive, with reference to a published algorithm (Pediatric Medical Complexity Algorithm[23]), because progressive diseases would be expected to have increasing health care usage over time. In the case of multiple diagnosis codes associated with NI on a single hospital record, the child was considered to have progressive disease if any of the codes were associated with progressive disease. Code lists are available from the corresponding author (K.E.N.).

Other Comorbidities

Because children with more medical complexity would be expected to have higher health care use, all hospital records from birth to time 0 were evaluated for diagnosis codes associated with other CCCs. Using both diagnosis and procedure codes, we also assessed for the presence of other medical technology at the time of feeding tube placement. Code lists are available (unpublished observations).

Demographic Characteristics

Because lower income and rural residency may be associated with challenges accessing the health care system, we evaluated for these characteristics in the cohort. We obtained the year and postal code from the hospital record that included feeding tube placement. The postal code was then linked with census data to determine neighborhood income quintile on the basis of the smallest census areas with aggregate data (400–700 people). Census data are

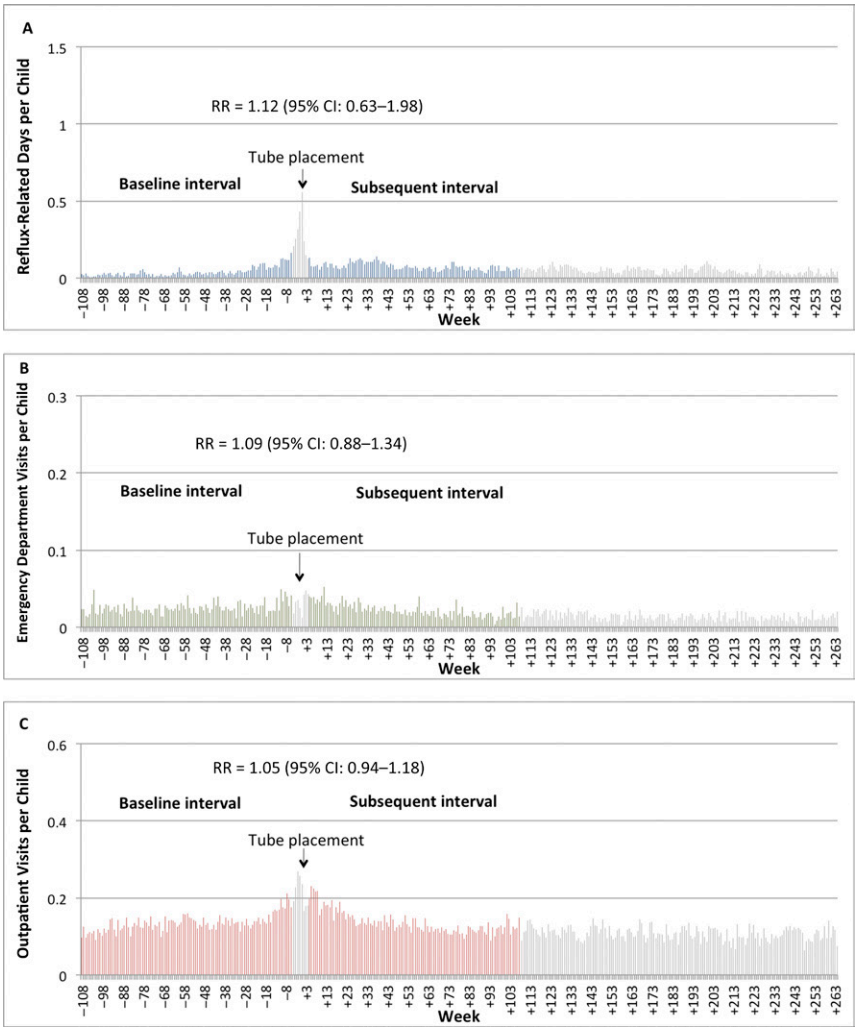
suppressed for areas with residential instability, which are typically low-income urban areas, so children with missing data were assumed to be part of the lowest income quintile. Rurality was defined by using postal codes according to the Rurality Index of Ontario; each community in Ontario has a score of 0 to 100 on the basis of markers, including population size, distance to nearest referral center, and availability of general practitioners. Scores ≥40 on the index were considered rural. Because most people in Ontario live in urban areas, children with missing rurality data were assumed to be "not rural."

Immigration Status

Immigration status was determined by the presence of either the child's or the child's birth mother's encoded identifier in the Citizenship and Immigration Canada database. If neither the child nor the mother appeared in the database, they were categorized as "not immigrant." We further delineated 3 types of immigrants: refugee, nonrefugee, new immigrant (either a child or mother with a date of immigration to Canada occurring <3 years before feeding tube placement), and nonrefugee, not new immigrant.

Provider Type

In the Canadian system, most children are cared for by general practitioners or family doctors, particularly outside major urban centers. Having a primary care pediatrician may be associated with more complex care needs.[27] Rostering data and physician billing records were used to determine primary care provider type. In primary care reform models, family medicine patients are rostered under specific physicians[28]; rostered children were assigned the provider type of that physician. For children who were not rostered, the algorithm evaluated primary care billing codes extracted between the child's birth and the date of the procedure. The child was assigned the specialty of the physician who provided the most primary care on the basis of billed fee codes. Fee codes were assessed hierarchically to make this determination, with most weight given to the codes for visits including anticipatory guidance and vaccines and then all other primary care visits.



SUPPLEMENTAL FIGURE 4
Bamboo plots. A, Reflux-related hospital days. B, Emergency department visits without admission. C, Outpatient visits.

ANTIREFLUX SURGERY CODES

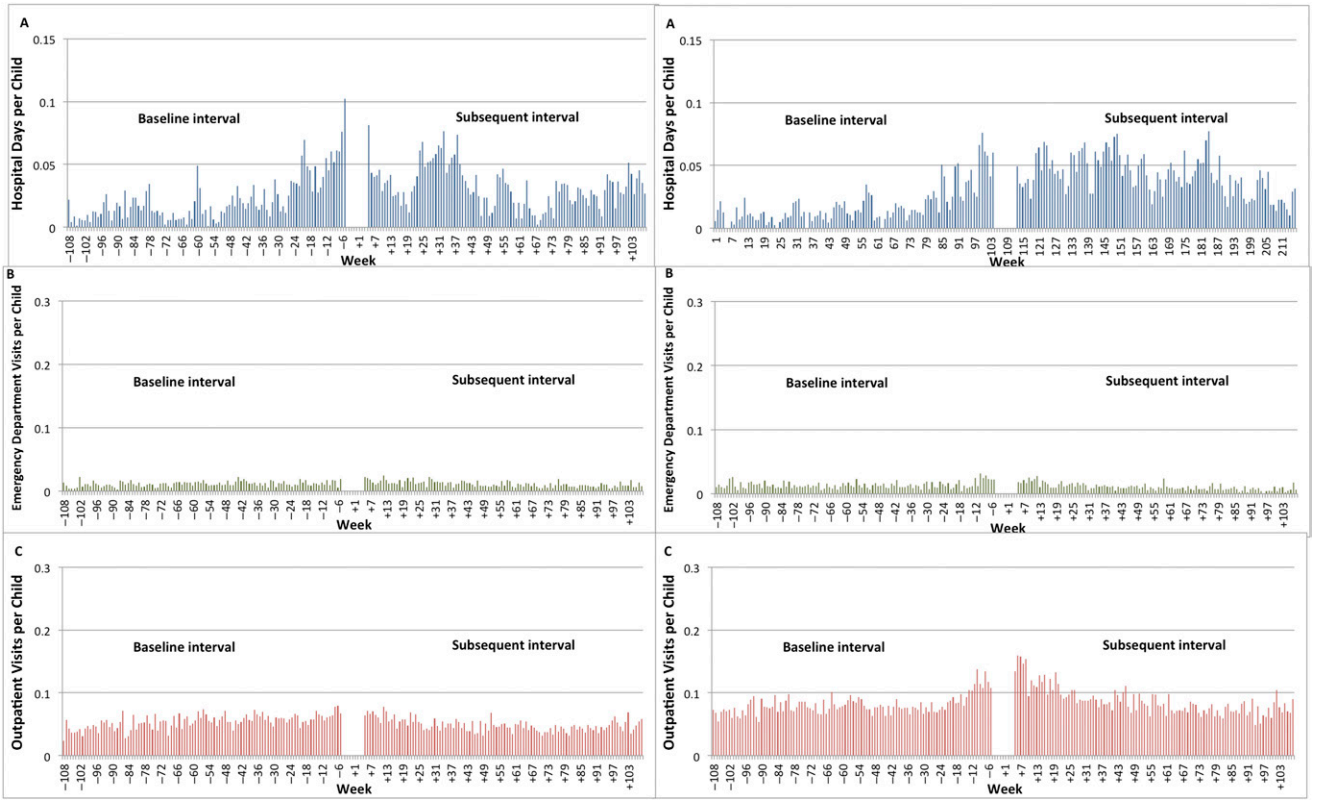
Hospital procedure codes <2002: 54.76, 56.59, and 65.7.

Hospital procedure code ≥2002: 1.NA.80.

Physician billing code: SO97.

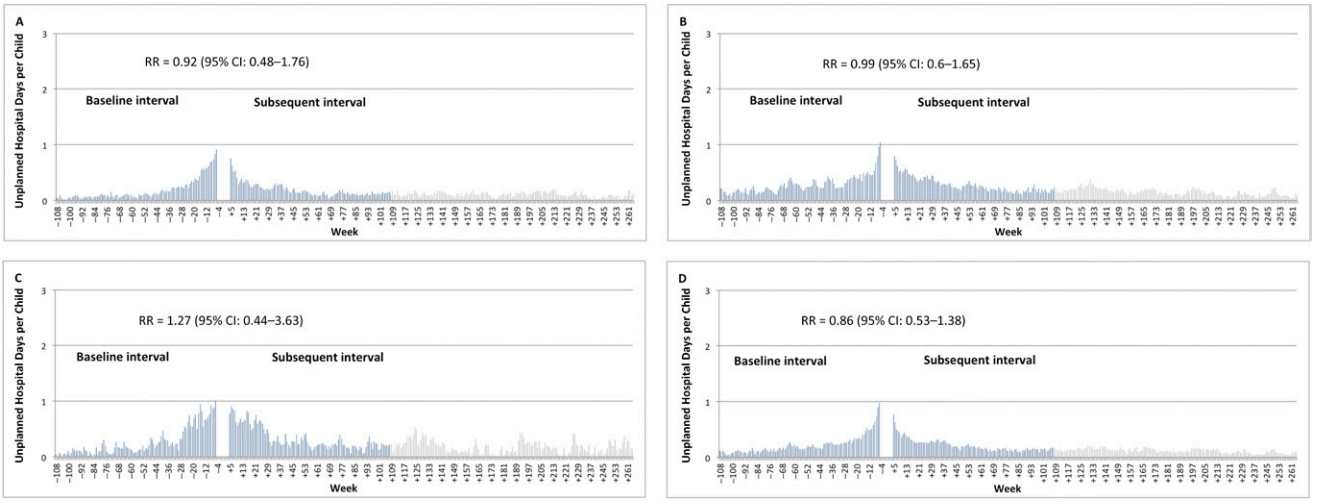
REFLUX-RELATED DIAGNOSIS CODES

International Classification of Diseases, Ninth Revision: 530.1, 530.8, 507.0, 507.1, 507.8, 480.0, 480.1, 480.2, 480.8, 480.9, 481, 481.0, 482.0, 482.1, 482.2, 482.3, 482.4,



SUPPLEMENTAL FIGURE 5

Bamboo plots. A, Reflux-related hospital days for children with (n = 404) and without (n = 544) antireflux procedures. B, Emergency department visits for children with (n = 404) and without (n = 544) antireflux procedures. C, Outpatient visits for children with (n = 404) and without (n = 544) antireflux procedures.

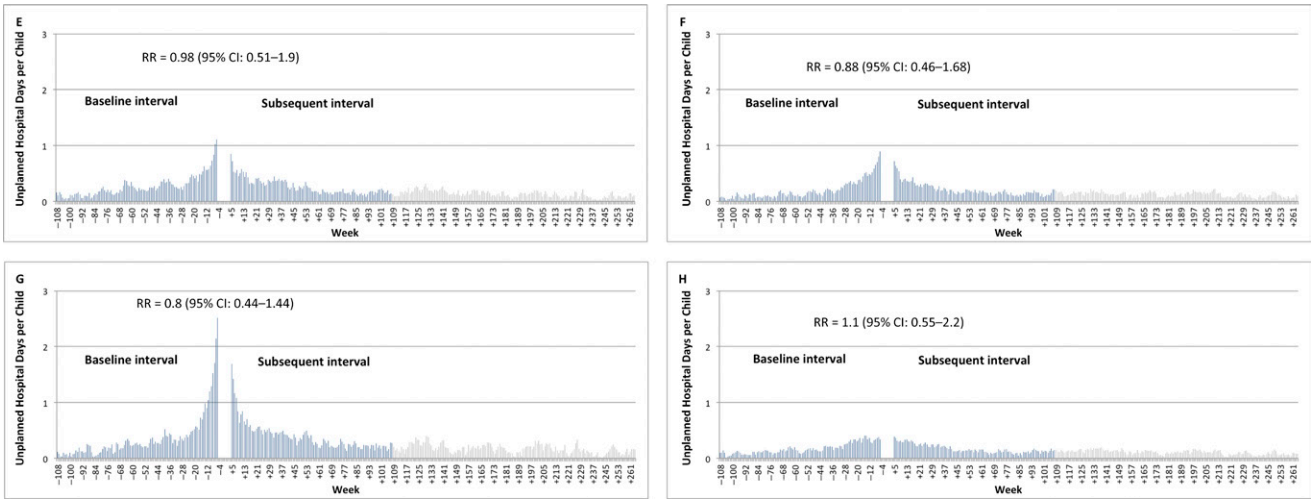


SUPPLEMENTAL FIGURE 6

Bamboo plots for days of unplanned hospitalization per child for each week for subgroups. A, Children ≥4 years old at the time of feeding tube placement (n = 480). B, Children <4 years old at the time of feeding tube placement (n = 468). C, Progressive NI (n = 156). D, Nonprogressive NI (n = 792).

482.8, 482.9, 483, 484.1, 484.3, 484.5, 484.6, 484.7, 484.8, 485, 485.0, 486, 486.0, 493.0, 493.00, 493.01, 493.1, 493.10, 493.11, 493.20, 493.21, 493.9, 493.90, 493.91, and 518.81.

International Classification of Diseases, 10th Revision: J15.2, J15.3, J15.4, J15.5, J15.6, J15.7, J15.8, J15.9, J16.0, J16.8, J17.0, J17.1, J17.2, J17.3, J17.8, J18.0, J18.1, J18.2, J18.8, J18.9, J45.00, J45.01, J45.10, J45.11, J45.80,



SUPPLEMENTAL FIGURE 7

Bamboo plots for days of unplanned hospitalization per child for each week for subgroups (continued). E, With antireflux procedure (n = 404). F, Without antireflux procedure (n = 544). G, Inpatient at tube placement (n = 279). H, Outpatient at tube placement (n = 669). The RRs were generated with a negative binomial GEE model, adjusting for time and age. The induction period is not shown on this graph, and the gray lines are used to indicate follow-up time not included in the exposure-crossover analysis.

SUPPLEMENTAL TABLE 4 Most Common Diagnoses in Each NI Category

	n (%)
Cerebral palsy (*n* = 467)	
Cerebral palsy, unspecified	254 (54.4)
Spastic quadriplegic cerebral palsy	126 (27.0)
Other cerebral palsy	47 (10.1)
Central nervous system degeneration and diseases (*n* = 189)	
Degenerative disease of nervous system	46 (24)
Obstructive hydrocephalus	36 (19)
Rett syndrome	24 (13)
Brain and spinal cord malformations (*n* = 178)	
Reduction deformity, brain	43 (24)
Congenital malformation of nervous system	28 (16)
Spina bifida	18 (10)
Muscular dystrophy, myopathies, and movement disorders (*n* = 70)	
Other mitochondrial myopathy	13 (19)
Muscular dystrophy	13 (19)
Myotonic disorders	10 (14)
Epilepsy (*n* = 55)	
Epilepsy, unspecified, intractable	26 (47)
Other generalized idiopathic epilepsy, intractable	11 (20)
Generalized idiopathic epilepsy and epileptic syndromes	10 (18)
Other disorders of central nervous system (*n* = 45)	
Anoxic brain damage	16 (36)
Quadriplegia, unspecified	<6
Cerebral artery occlusion, unspecified	<6
Genetic and metabolic (*n* = 37)	
Down syndrome	14 (38)
Prader-Willi syndrome	13 (35)
Lipidoses	<6
Malignancy (*n* = 22)	
Malignant neoplasm of the cerebellum, unspecified	9 (41)
Malignant neoplasm of the brain, unspecified	6 (27)
Malignant neoplasm of the brain stem	<6

Percentages are based on the total number of children with diagnoses in each category. Children with diagnoses in multiple categories contribute to the count for each of those categories.

J45.81, J45.90, J45.91, J69.0, J69.1, J69.8, K20, K21.0, K21.9, and J96.0.

SUPPLEMENTAL TABLE 5 Top 3 Diagnoses in Each Nonneurologic CCC Category

	n (%)
Neonatal (*n* = 220)	
Bronchopulmonary dysplasia	57 (25.9)
Newborn asphyxia, unspecified	56 (25.5)
Newborn intraventricular hemorrhage	22 (10)
Genetic (*n* = 144)	
Other chromosomal and congenital anomalies	43 (30)
Scoliosis	34 (24)
Congenital diaphragmatic hernia	17 (12)
Cardiologic (*n* = 104)	
Bradycardia, unspecified	24 (23)
Pulmonary artery anomaly	17 (12)
Tetralogy of Fallot	<6
Metabolic (*n* = 99)	
Metabolic disorder, unspecified	21 (21)
Diabetes insipidus	14 (14)
Syndrome of inappropriate secretion of antidiuretic hormone	12 (12)
Pulmonary (*n* = 62)	
Congenital laryngomalacia and tracheomalacia	35 (57)
Hypoplasia and dysplasia of lung	10 (16)
Primary atelectasis of newborn	6 (10)
Hematologic (*n* = 44)	
Neutropenia	15 (34)
Aplastic anemia	8 (18)
Thalassemia	7 (16)
Neoplastic (*n* = 42)	
Secondary malignant neoplasm of bone and bone marrow	8 (19)
Acute lymphoblastic leukemia	<6
Neoplasm of uncertain or unknown behavior of brain	<6
Gastrointestinal (*n* = 39)	
Atresia of esophagus without fistula	6 (15)
Congenital malformations of intestinal fixation	<6
Atresia of large intestine	<6
Renal (*n* = 37)	
Atony of bladder	9 (24)
Renal agenesis and other specified congenital malformations of the kidney	9 (24)
Chronic kidney disease, unspecified	<6
Miscellaneous (*n* <6)	
Complication of transplanted organ and transplant status not otherwise classified	—
Rejection of other transplanted tissue	—

Percentages are calculated on the basis of number of diagnoses within each category. Children with diagnoses in multiple categories contribute to the count for each of those categories. —, not applicable.

SUPPLEMENTAL TABLE 6 Most Common Primary Reasons for Admission in the Baseline and Subsequent Intervals

Baseline Interval (n = 2388)	n (%)	Subsequent Interval (n = 2228)	n (%)
Seizures and epilepsies	395 (16.5)	Pneumonia (unspecified, bacterial, and viral)	309 (13.9)
Pneumonia (unspecified, bacterial, and viral)	255 (10.7)	Aspiration pneumonia	298 (13.4)
Aspiration pneumonia	164 (6.9)	Seizures and epilepsies	222 (10)
Bronchiolitis	82 (3.4)	Acute upper respiratory infection	87 (3.9)
Failure to thrive	71 (3)	Vomiting	60 (2.7)
Asthma	68 (2.8)	Asthma	54 (2.4)
Acute upper respiratory infection	62 (2.6)	Gastroenteritis	51 (2.3)
Dehydration	60 (2.5)	Gastrostomy malfunction	49 (2.2)
Feeding problem	58 (2.4)	Fever	42 (1.9)
Gastroenteritis	53 (2.2)	Neutropenia	42 (1.9)
Vomiting	47 (2)	Viral infection	39 (1.8)
Viral infection	46 (1.9)	Bronchiolitis	34 (1.5)
Respiratory abnormality	43 (1.8)	Dehydration	34 (1.5)
Urinary tract infection	43 (1.8)	Shortness of breath	33 (1.5)
Influenza	29 (1.2)	Chemotherapy	32 (1.4)
Gastrostomy malfunction	27 (1.1)	Feeding problem	32 (1.4)
Malignant neoplasm of cerebellum	25 (1)	Urinary tract infection	31 (1.4)
Fever	24 (1)	Sepsis	30 (1.3)
Esophagitis	21 (0.9)	Influenza	29 (1.3)
Neutropenia	19 (0.8)	Mechanical complication of intracranial shunt	20 (0.9)

SUPPLEMENTAL TABLE 7 Results of Sensitivity Analyses Testing for Survivor Bias, Era Effect, and Model Robustness

	N	RR (95% CI)
Survivors	838	0.64 (0.44–0.94)
Years 1993–2000	165	0.69 (0.32–1.5)
Years 2001–2005	275	1.02 (0.46–2.29)
Years 2006–2010	277	0.91 (0.34–2.42)
Years 2011–2015	231	1.01 (0.43–2.36)
Excluding outliers	936	0.91 (0.56–1.49)
Short induction (±2 wk)	948	0.89 (0.59–1.36)
Long induction (±6 wk)	948	0.92 (0.57–1.49)
Double induction (±8 wk)	948	0.92 (0.56–1.49
Triple induction (±12 wk)	948	0.92 (0.55–1.51)

Adjusted RRs were calculated with a negative binomial GEE adjusting for time and age at procedure.

SUPPLEMENTAL TABLE 8 Most Common Primary Reasons for Emergency Department Visits Without Admission in the Baseline and Subsequent Intervals

Baseline Interval (n = 2376)	n (%)	Subsequent Interval (n = 2063)	n (%)
Seizures and epilepsies	202 (8.5)	Gastrostomy malfunction	212 (10.3)
Pneumonia (unspecified, bacterial, and viral)	142 (6)	Pneumonia (unspecified, bacterial, and viral)	147 (7.1)
Pharyngitis	119 (5)	Seizures and epilepsies	123 (6)
Upper respiratory infection	118 (5)	Viral infection	106 (5.1)
Viral infection	108 (4.6)	Vomiting	104 (5)
Otitis media	103 (4.3)	Upper respiratory infection	99 (4.8)
Vomiting	95 (4)	Fever	64 (3.1)
Gastroenteritis	83 (3.5)	Pharyngitis	60 (2.9)
Coma and stupor	74 (3.1)	Gastroenteritis	55 (2.7)
Bronchitis	68 (2.9)	Otitis media	53 (2.6)

SUPPLEMENTAL TABLE 9 Procedure, Diagnosis, and Billing Codes

Feeding Tube Codes	Procedure	Tube Type
Hospital procedure codes		
<2002		
552	Permanent gastrostomy	GT
5839	Other enterostomy, not elsewhere classified	Unclear
5510	Temporary gastrostomy	GT
Hospital procedure codes		
≥2002		
1NF53BABC	Implantation of internal device, stomach using endoscopic per orifice approach of pneumatic balloon	GT
1NF53BATS	Implantation of internal device, stomach using endoscopic per orifice approach and tube NOS	GT
1NF53BTQB	Implantation of internal device, stomach of (gastric) valved tube using per orifice endoscopic approach with percutaneous incision	GT
1NF53BTTS	Implantation of internal device, stomach of (gastric) tube using per orifice endoscopic approach with percutaneous incision	GT
1NF53DAQB	Implantation of internal device, stomach of (gastric) valved tube using endoscopic (laparoscopic) approach	GT
1NF53DATS	Implantation of internal device, stomach of (gastric) tube using endoscopic (laparoscopic) approach	GT
1NF53HATS	Implantation of internal device, stomach of (gastric) tube using percutaneous approach	GT
1NF53LAQB	Implantation of internal device, stomach of (gastric) valved tube using open (laparotomy) approach	GT
1NF53LATS	Implantation of internal device, stomach of (gastric) tube using open (laparotomy) approach	GT
10W35CAD1	Pharmacotherapy, surgically constructed sites in digestive and biliary tract using per orifice approach and anti-infective irrigating solution	Unclear
10W35CAD2	Drainage, stomach using percutaneous needle approach [injection] and tube NOS	GT
10W35CAD3	Pharmacotherapy, surgically constructed sites in digestive and biliary tract using per orifice approach and other irrigating solution	Unclear
10W35HAD1	Pharmacotherapy, surgically constructed sites in digestive and biliary tract using percutaneous needle approach (injection) and anti-infective irrigating solution	Unclear
10W35HAD2	Pharmacotherapy, surgically constructed sites in digestive and biliary tract using percutaneous needle approach (injection) and salt irrigating solution	Unclear
10W35HAD3	Pharmacotherapy, surgically constructed sites in digestive and biliary tract using percutaneous needle approach (injection) and other irrigating solution	Unclear
10W12ZZ	Therapy, surgically constructed sites in digestive and biliary tract using technique NEC	Unclear
1NK77EM	Bypass with exteriorization, small intestine endoscopic (laparoscopic)-approach feeding enterostomy (eg, jejunostomy)	GJT
1NK77RQ	Bypass with exteriorization, small intestine open-approach feeding enterostomy (eg, jejunostomy)	GJT
1NK53DATS	Implantation of internal device, small intestine of feeding tube (jejunal) using endoscopic (laparoscopic) approach	GJT
1NK53LATS	Implantation of internal device, small intestine of feeding tube (jejunal) using open approach	GJT
1NP54JATS	Management of internal device, small with large intestine using external approach and tube NOS	GJT
1NF52HATS	Drainage, stomach using percutaneous needle approach (injection) and tube NOS	GT
1NF53BATS	Implantation of internal device, stomach using endoscopic per orifice approach and tube NOS	GT
1NF54JAHG	Management of internal device, stomach using external approach and cystostomy tube	GT
1NF53BABC	Implantation of internal device, stomach using endoscopic per orifice approach of pneumatic balloon	GT
Physician billing codes		
J055	Percutaneous gastrostomy	GT
S118	Gastrostomy	GT
Z532	Percutaneous endoscopic gastrostomy	GT
S134	Gastroduodenostomy or gastrojejunostomy	GJT
J063	Percutaneous jejunostomy	GJT
J064	Exchange of drainage tubes	Unclear
Z540	Manipulation and/or intubation of small bowel	GJT
J055	Percutaneous gastrostomy	GT

NEC, not elsewhere classified; NOS, not otherwise specified.

SUPPLEMENTAL TABLE 10 NI Codes and Progressive Disease Status

	ICD-9	ICD-10	Progressive
Brain and spinal cord malformations			
Anencephalus	740.0	—	Yes
Craniorachischisis	740.1	—	Yes
Iniencephaly	740.2	—	Yes
Encephalocele	742.0	—	No
Microcephalus	742.1	—	No
Reduction deformities, brain	742.2	—	No
Congenital hydrocephalus	742.3	—	No
Brain anomaly NEC	742.4	—	No
Nervous system anomaly NEC	742.8	—	No
Nervous system anomaly NOS	742.9	—	No
Spina bifida with hydrocephalus NOS	741.00	—	No
Spina bifida with hydrocephalus-cervical	741.01	—	No
Spina bifida with hydrocephalus-dorsal	741.02	—	No
Spina bifida with hydrocephalus-lumbar	741.03	—	No
Spina bifida	741.90	—	No
Spina bifida-cervical	741.91	—	No
Spina bifida-dorsal	741.92	—	No
Spina bifida-lumbar	741.93	—	No
Diastematomyelia	742.51	—	No
Hydromyelia	742.53	—	No
Spinal cord anomaly NEC	742.59	—	No
Familial dysautonomia (Riley-Day)	—	G90.1	No
Anencephaly	—	Q00.0	Yes
Craniorachischisis	—	Q00.1	Yes
Iniencephaly	—	Q00.2	Yes
Frontal encephalocele	—	Q01.0	No
Nasofrontal encephalocele	—	Q01.1	No
Occipital encephalocele	—	Q01.2	No
Encephalocele of other sites	—	Q01.8	No
Encephalocele, unspecified	—	Q01.9	No
Microcephaly	—	Q02	No
Malformations of aqueduct of Sylvius	—	Q03.0	No
Atresia of foramina of Magendie and Luschka	—	Q03.1	No
Other congenital hydrocephalus	—	Q03.8	No
Congenital hydrocephalus, unspecified	—	Q03.9	No
Congenital malformations of corpus callosum	—	Q04.0	No
Arhinencephaly	—	Q04.1	Yes
Holoprosencephaly	—	Q04.2	Yes
Other reduction deformities of brain	—	Q04.3	No
Septo-optic dysplasia	—	Q04.4	No
Megalencephaly	—	Q04.5	Yes
Congenital cerebral cysts	—	Q04.6	No
Other specified congenital malformations of brain	—	Q04.8	No
Congenital malformation of brain, unspecified	—	Q04.9	No
Cervical spina bifida with hydrocephalus	—	Q05.0	No
Thoracic spina bifida with hydrocephalus	—	Q05.1	No
Lumbar spina bifida with hydrocephalus	—	Q05.2	No
Sacral spina bifida with hydrocephalus	—	Q05.3	No
Unspecified spina bifida with hydrocephalus	—	Q05.4	No
Cervical spina bifida without hydrocephalus	—	Q05.5	No
Thoracic spina bifida without hydrocephalus	—	Q05.6	No
Lumbar spina bifida without hydrocephalus	—	Q05.7	No
Sacral spina bifida without hydrocephalus	—	Q05.8	No
Spina bifida, unspecified	—	Q05.9	No
Amyelia	—	Q06.0	Yes
Hypoplasia and dysplasia of spinal cord	—	Q06.1	No
Diastematomyelia	—	Q06.2	No
Other congenital cauda equina malformations	—	Q06.3	No
Hydromyelia	—	Q06.4	No
Other specified congenital malformations of spinal cord	—	Q06.8	No
Congenital malformation of spinal cord, unspecified	—	Q06.9	No

TABLE 10 Continued

	ICD-9	ICD-10	Progressive
Arnold-Chiari syndrome	—	Q07.0	No
Other specified congenital malformations of nervous system	—	Q07.8	No
Congenital malformation of nervous system, unspecified	—	Q07.9	No
Central nervous system degeneration and diseases			
Leukodystrophy	330.0	—	Yes
Cerebral lipidoses	330.1	—	Yes
Cerebral degeneration in lipidosis	330.2	—	Yes
Cerebral degeneration of childhood in other disease	330.3	—	Yes
Cerebral degeneration in childhood NEC	330.8	—	Yes
Cerebral degeneration in child NOS	330.9	—	Yes
Pick's disease	331.11	—	Yes
Obstructive hydrocephalus	331.4	—	No
Cerebral degeneration NOS	331.9	—	Yes
Myoclonus	333.2	—	No
Friedreich's ataxia	334.0	—	Yes
Hereditary spastic paraplegia	334.1	—	Yes
Primary cerebellar degeneration	334.2	—	Yes
Cerebellar ataxia NEC	334.3	—	No
Cerebellar ataxia in other disease	334.4	—	Yes
Spinocerebellar disease NEC	334.8	—	No
Spinocerebellar disease NOS	334.9	—	No
Werdnig-Hoffmann disease	335.0	—	Yes
Anterior horn cell disease NEC	335.8	—	Yes
Anterior horn cell disease NOS	335.9	—	Yes
Vascular myelopathies	336.1	—	No
Myelopathy NEC	336.8	—	No
Autonomic nerve disease NEC	337.9	—	No
Cerebral degeneration NEC	331.89	—	Yes
Spinal muscular atrophy NOS	335.10	—	Yes
Kugelberg-Welander disease	335.11	—	Yes
Spinal muscular atrophy NEC	335.19	—	Yes
Amyotrophic sclerosis	335.20	—	Yes
Progressive muscular atrophy	335.21	—	Yes
Progressive bulbar palsy	335.22	—	Yes
Pseudobulbar palsy	335.23	—	Yes
Primary lateral sclerosis	335.24	—	Yes
Motor neuron disease NEC	335.29	—	Yes
GM2 gangliosidosis	—	E75.0	Yes
Other gangliosidosis	—	E75.1	Yes
Other sphingolipidosis	—	E75.2	Yes
Neuronal ceroid lipofuscinosis	—	E75.4	Yes
Rett's syndrome	—	F84.2	No
Early-onset cerebellar ataxia	—	G11.1	Yes
Late-onset cerebellar ataxia	—	G11.2	Yes
Cerebellar ataxia with defective DNA repair	—	G11.3	Yes
Hereditary spastic paraplegia	—	G11.4	Yes
Other hereditary ataxias	—	G11.8	Yes
Hereditary ataxia, unspecified	—	G11.9	Yes
Infantile spinal muscular atrophy, type I (Werdnig-Hoffman)	—	G12.0	Yes
Other inherited spinal muscular atrophy	—	G12.1	Yes
Motor neuron disease	—	G12.2	Yes
Other spinal muscular atrophies and related syndromes	—	G12.8	Yes
Spinal muscular atrophy, unspecified	—	G12.9	Yes
Myoclonus	—	G25.3	No
Circumscribed brain atrophy	—	G31.0	Yes
Other specified degenerative diseases of nervous system	—	G31.8	Yes
Degenerative disease of nervous system, unspecified	—	G31.9	Yes
Other specified degenerative disorders of nervous system in diseases classified elsewhere	—	G32.8	Yes
Disorder of autonomic nervous system, unspecified	—	G90.9	No
Obstructive hydrocephalus	—	G91.1	No
Metabolic encephalopathy	—	G93.80	No
Neurologically determined death	—	G93.81	Yes
Other specified disorders of brain	—	G93.88	No

TABLE 10 Continued

	ICD-9	ICD-10	Progressive
Disorder of brain, unspecified	—	G93.9	No
Hydrocephalus in infectious and parasitic diseases classified elsewhere	—	G94.0	No
Hydrocephalus in neoplastic disease	—	G94.1	No
Hydrocephalus in other diseases classified elsewhere	—	G94.2	No
Tuberous sclerosis	—	Q85.1	No
Central nervous system malignancy			
Malignant neoplasm of cerebrum	191.0	—	Yes
Malignant neoplasm of temporal lobe	191.2	—	Yes
Malignant neoplasm of parietal lobe	191.3	—	Yes
Malignant neoplasm of occipital lobe	191.4	—	Yes
Malignant neoplasm of cerebrum ventricle	191.5	—	Yes
Malignant neoplasm of cerebellum NOS	191.6	—	Yes
Malignant neoplasm of brain stem	191.7	—	Yes
Malignant neoplasm of brain NEC	191.8	—	Yes
Malignant neoplasm of brain NOS	191.9	—	Yes
Malignant neoplasm of cranial nerves	192.0	—	Yes
Malignant neoplasm of cerebral meninges	192.1	—	Yes
Secondary malignant neoplasm of brain and spinal cord	198.3	—	Yes
Malignant neoplasm of cerebral meninges	—	C70.0	Yes
Malignant neoplasm of meninges, unspecified	—	C70.9	Yes
Malignant neoplasm of cerebrum, except lobes and ventricles	—	C71.0	Yes
Malignant neoplasm of temporal lobe	—	C71.2	Yes
Malignant neoplasm of parietal lobe	—	C71.3	Yes
Malignant neoplasm of occipital lobe	—	C71.4	Yes
Malignant neoplasm of cerebral ventricle	—	C71.5	Yes
Malignant neoplasm of cerebellum	—	C71.6	Yes
Malignant neoplasm of brain stem	—	C71.7	Yes
Overlapping malignant lesion of brain	—	C71.8	Yes
Malignant neoplasm of brain unspecified	—	C71.9	Yes
Malignant neoplasm of olfactory nerve	—	C72.2	Yes
Malignant neoplasm of optic nerve	—	C72.3	Yes
Malignant neoplasm of acoustic nerve	—	C72.4	Yes
Malignant neoplasm of other and unspecified cranial nerves	—	C72.5	Yes
Secondary malignant neoplasm of brain and cerebral meninges	—	C79.3	Yes
Cerebral palsy			
Congenital diplegia	343.0	—	No
Congenital hemiplegia	343.1	—	No
Congenital quadriplegia	343.2	—	No
Congenital monoplegia	343.3	—	No
Infantile hemiplegia	343.4	—	No
Cerebral palsy NEC	343.8	—	No
Cerebral palsy NOS	343.9	—	No
Spastic quadriplegic cerebral palsy	—	G80.0	No
Spastic diplegic cerebral palsy	—	G80.1	No
Spastic hemiplegic cerebral palsy	—	G80.2	No
Ataxic cerebral palsy	—	G80.4	No
Other cerebral palsy	—	G80.8	No
Cerebral palsy, unspecified	—	G80.9	No
Epilepsy			
Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with simple partial seizures, intractable	—	G40.11	No
Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with complex partial seizures, intractable	—	G40.21	No
Generalized idiopathic epilepsy and epileptic syndromes	—	G40.30	No
Generalized idiopathic epilepsy and epileptic syndromes, intractable	—	G40.31	No
Other generalized idiopathic epilepsy and epileptic syndromes, intractable	—	G40.41	No
Other epilepsy, not stated as intractable	—	G40.80	No
Epilepsy, unspecified, intractable	—	G40.91	No
Genetic and metabolic			
Down syndrome	758.0	—	No
Patau syndrome	758.1	—	Yes
Edwards syndrome	758.2	—	Yes
Prader-Willi syndrome	759.81	—	No

TABLE 10 Continued

	ICD-9	ICD-10	Progressive
Phenylketonuria	270.1	—	Yes
Aromatic amino-acid metabolism NEC	270.2	—	Yes
Lipidoses	272.7	—	Yes
Purine or pyrimidine disease NEC	277.2	—	Yes
Congenital malformation syndromes predominantly associated with short stature	—	Q87.1	No
Down syndrome, unspecified	—	Q90.9	No
Edwards syndrome, unspecified	—	Q91.3	Yes
Trisomy 13, meiotic nondisjunction	—	Q91.4	Yes
Patau syndrome, unspecified	—	Q91.7	Yes
Classic phenylketonuria	—	E70.0	Yes
Disorders of tyrosine metabolism	—	E70.2	Yes
Albinism	—	E70.3	No
Other disorders of aromatic amino-acid metabolism	—	E70.8	Yes
Sphingolipidosis, unspecified	—	E75.3	Yes
Other lipid storage disorders	—	E75.5	Yes
Lipid storage disorder, unspecified	—	E75.6	Yes
Defects in posttranslational modification of lysosomal enzymes	—	E77.0	Yes
Lesch-Nyhan syndrome	—	E79.1	Yes
Other disorders of purine and pyrimidine metabolism	—	E79.8	Yes
Other paralytic strabismus	—	H49.8	No
Muscular dystrophy, myopathies and movement disorders			
Paralysis agitans	332.0	—	No
Secondary parkinsonism	332.1	—	No
Degeneration basal ganglia NEC	333.0	—	Yes
Chorea NEC	333.5	—	Yes
Symptom torsion dystonia	333.7	—	No
Congenital hereditary muscular dystrophy	359.0	—	Yes
Hereditary progressive muscular dystrophy	359.1	—	Yes
Myotonic disorders	359.2	—	Yes
Familial periodic paralysis	359.3	—	No
Huntington disease	—	G10	Yes
Parkinson disease	—	G20	Yes
Malignant neuroleptic syndrome	—	G21.0	No
Other drug-induced secondary parkinsonism	—	G21.1	No
Other secondary parkinsonism	—	G21.8	No
Hallervorden-Spatz disease	—	G23.0	Yes
Progressive supranuclear ophthalmoplegia (Steele-Richardson-Olszewski)	—	G23.1	Yes
Striatonigral degeneration	—	G23.2	Yes
Other specified degenerative diseases of basal ganglia	—	G23.8	Yes
Other dystonia	—	G24.8	No
Drug-induced chorea	—	G25.4	No
Other chorea	—	G25.5	No
Other specified extrapyramidal and movement disorders	—	G25.8	No
Extrapyramidal and movement disorder, unspecified	—	G25.9	No
Dyskinetic cerebral palsy	—	G80.3	No
Muscular dystrophy	—	G71.0	Yes
Myotonic disorders	—	G71.1	No
Congenital myopathies	—	G71.2	Yes
Mitochondrial myopathy, not elsewhere classified	—	G71.3	Yes
Other primary disorders of muscles	—	G71.8	No
Primary disorder of muscle, unspecified	—	G71.9	No
Drug-induced myopathy	—	G72.0	No
Myopathy due to other toxic agents	—	G72.2	No
Periodic paralysis	—	G72.3	No
Inflammatory myopathy, not elsewhere classified	—	G72.4	No
Other specified myopathies	—	G72.8	No
Myopathy, unspecified	—	G72.9	No
Other disorders of central nervous system			
Cerebral thrombosis	434.0	—	No
Cerebral artery occlusion NOS	434.9	—	No
CNS demyelination NEC	341.8	—	Yes
Hemiplegia NOS	342.9	—	No
Quadriplegia NOS	344.0	—	No

TABLE 10 Continued

	ICD-9	ICD-10	Progressive
Paralysis NOS	344.9	—	No
Anoxic brain damage	348.1	—	No
Compression of brain	348.4	—	No
Neurogenic bladder	344.61	—	No
Severe birth asphyxia	768.5	—	No
Congenital rubella	771.0	—	No
Congenital cytomegalovirus infection	771.1	—	No
Newborn kernicterus due to isoimmunization	773.4	—	No
Newborn kernicterus	774.7	—	No
Moderate mental retardation	318.0	—	No
Severe mental retardation	318.1	—	No
Profound mental retardation	318.2	—	No
Moderate mental retardation with the statement of no or minimal impairment of behavior	—	F71.0	No
Moderate mental retardation, significant impairment of behavior requiring attention or treatment	—	F71.1	No
Moderate mental retardation, other impairments of behavior	—	F71.8	No
Moderate mental retardation without mention of impairment of behavior	—	F71.9	No
Severe mental retardation with the statement of no, or minimal, impairment of behavior	—	F72.0	No
Severe mental retardation, significant impairment of behavior requiring attention or treatment	—	F72.1	No
Severe mental retardation, other impairments of behavior	—	F72.8	No
Severe mental retardation without mention of impairment of behavior	—	F72.9	No
Profound mental retardation with the statement of no, or minimal, impairment of behavior	—	F73.0	No
Profound mental retardation, significant impairment of behavior requiring attention or treatment	—	F73.1	No
Profound mental retardation, other impairments of behavior	—	F73.8	No
Profound mental retardation without mention of impairment of behavior	—	F73.9	No
Cerebral infarction due to thrombosis of cerebral arteries	—	I63.3	No
Cerebral infarction due to unspecified occlusion or stenosis of cerebral arteries	—	I63.5	No
Central demyelination of corpus callosum	—	G37.1	Yes
Central pontine myelinolysis	—	G37.2	Yes
Other specified demyelinating diseases of central nervous system	—	G37.8	Yes
Hemiplegia of unspecified type of dominant side	—	G81.90	No
Quadriplegia, unspecified type, complete, at cervical spine level C1 to C4	—	G82.510	No
Quadriplegia, unspecified type, complete, at cervical spine level C5 to C7	—	G82.511	No
Quadriplegia, unspecified type, incomplete, at cervical spine level C1 to C4	—	G82.520	No
Quadriplegia, unspecified type, incomplete, at cervical spine level C5 to C7	—	G82.521	No
Cauda equina syndrome	—	G83.4	No
Locked-in state	—	G83.5	No
Paralytic syndrome, unspecified	—	G83.9	No
Anoxic brain damage, not elsewhere classified	—	G93.1	No
Compression of brain	—	G93.5	No
Subdural hemorrhage due to birth injury	—	P10.0	No
Cerebral hemorrhage due to birth injury	—	P10.1	No
Tentorial tear due to birth injury	—	P10.4	No
Congenital rubella syndrome	—	P35.0	No
Congenital cytomegalovirus infection	—	P35.1	No
Other intracranial (nontraumatic) hemorrhages of fetus and newborn	—	P52.8	No
Kernicterus due to isoimmunization	—	P57.0	No
Other specified kernicterus	—	P57.8	No

ICD-9, *International Classification of Diseases, Ninth Revision*; ICD-10, *International Classification of Diseases, 10th Revision*; NEC, not elsewhere classified; NOS, not otherwise specified; —, not applicable.

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