

# Tethered Cord Syndrome in the United States: Cluster Analysis of Presenting Current Anomalies and Associated Conditions

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# **Learning Objectives**

- 1) Describe the importance of identifying the top concurrent anomalies present in tethered cord patients in the United States
- 2) Discuss, in small groups, the most common concurrent anomalies for each body system

# Introduction

- Tethered cord syndrome(TCS) is a form of an occult spinal dysraphism which includes:

 Low lying conus, tight filum terminale, lipomeningomyelocele, split cord malformations, dermal sinus tracts, and dermoids

- Associated with musculoskeletal, neurological, and gastrointestinal abnormalities

## Methods

 Retrospective review of the prospectively collected Nationwide Inpatient Sample(NIS) database from 2003-2012.

- NIS supplied hospital- and year-adjusted weights allowed for:

 accurate assessment of incidence of TCS, cardiac and gastrointestinal(GI) and genitourinary(GU) anomalies

Body System Anomaly	Frequency			
Spine	3298 (24.48%)			
Syndrome	9.67%			
Cardiac	6.27%			
Urinary	5.37%			
Gastrointestinal	4.55%			
Nervous	3.16%			
Genital & Urinary	2.48%			
Bone	1.01%			
Neck	1.00%			
Face	0.68%			
Eye	0.44%			
Ear	0.40%			
Lung	0.36%			
Endocrine system	0.01%			

### Results

- N=13,470 patients w/ TCS diagnosis
- 40.7% have at least one additional anomaly

Secondary anomalies most frequently presented in the following systems:

- spine (24.48%), cardiac (6.27%), urinary (5.37%), GI (4.55%), nervous (3.16%), GU (2.48%), and bone (1.01%)
- 12.55% of TCS patients had more than one body system anomal

Of patients with more than one associated anomaly, the most common combination of body system anomalies were:

• GI and cardiac (4.55%), urinary and GI (4.26%), and urinary and cardiac (4.19%).

The most common spinal associations were:

 13.65% spina bifida, 0.39% Klippel-Feil, and 0.12% with torticollis

The most common neurological and musculoskeletal anomalies were:

 13.45% with any VACTERL association, 3.58% with Rubenstein-Taybi syndrome, 0.42% with hydromyelia, 0.16% with chromosome 22 defect/deletion

23.8% of TCS patients with microcephalus also had patent ductus arteriosus and 18.0% of TCS patients with reduction deformities of the brain presented with atrial septal defect

The most common specific anomalies were:

• spina bifida, large intestine atresia, Rubenstein-Taybi syndrome, and atrial and ventral septal defects

Anomaly Type	Neck	Bone	Nervous System	Genetic	Spine	Urinary Genital	Gastro- Intestinal	Cardiac
Cardiac	53	30	107	124	130	230	250	
Gastrointestinal Urinary &	31	20	42	138	98	234		250
Genital	21	29	59	120	151		234	230
Spinal	8	46	66	52		151	98	130
Syndrome	48	36	87	96	471	225	201	287
Genetic	12	8	26		52	120	138	124
Nervous system	31	11	-	26	66	59	42	107
Bone	5		11	8	46	29	20	30
Neck		5	31	12	8	21	31	53
Eye	6	2	15	3	8	8	7	16
Ear	4	3	7	9	7	17	7	20
Lung	4	13	9	8	6	13	8	16
Facial	10	2	19	26	4	25	22	44

# Conclusions

- This study provides a nationwide prospective on congenital anomalies and concurrent conditions present in the tethered cord syndrome patient population in the United States.
- 40.7% of TCS patients have at least one associated anomaly reported.
- The most common congenital anomalies studied were spina bifida, urogenital with or without cardiac septal defects, cytourethral anomaly or cystic kidney disease with or without large intestinal atresia.

#### References

 Healthcare Cost and Utilization Project (HCUP): Overview of the Nationwide Inpatient Sample (NIS). Agency for Healthcare Research and Quality;2014.
Healthcare Cost and Utilization Project (HCUP): 2010 Introduction to the Nationwide Inpatient Sample (NIS). Agency for Healthcare Research and Quality;2014.

Scoliosis Diagnosis	N	Percentage	
No scoliosis	12173	90.37%	
Idiopathic Scoliosis	751	5.58%	
Congenital Scoliosis	227	1.69%	
Scoliosis not elsewhere classified	156	1.16%	
Postural Kyphosis	51	0.38%	
Scoliosis Associated with Other Conditions	42	0.31%	
Progressive Infantile Idiopathic Scoliosis	18	0.13%	
Thoracic Congenital Scoliosis	17	0.13%	
Kyphosis not elsewhere classified	10	0.07%	
Kyphosis Associated with Other Conditions	10	0.07%	
Post-laminectomy Kyphosis	1	0.01%	
Curvature not elsewhere classified	1	0.01%	
Lordosis Associated with Other Conditions	1	0.01%	
Other Spine Curves	1	0.01%	
Unspecified Curvature	1	0.01%	