

Testicular Torsion in Patients With Intellectual and Developmental Disabilities

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Abstract

Patients with intellectual/developmental disabilities are at risk of delayed diagnosis of testicular torsion due to their inability to effectively communicate their symptoms. We identified males ages 1 to 18 years with testicular torsion between January 1, 2015 and December 31, 2020, focusing on patients with intellectual and/or developmental disabilities. Of the 140 patients with testicular torsion, 5 (3.6%) patients exhibited intellectual/developmental disabilities with an inability to effectively verbalize testicular/groin/scrotal pain. The patients with intellectual/developmental disabilities underwent more orchiectomies (5/5, 100%, $P=.009$) and had a longer duration of symptoms (median = 48 hours, $P=.047$) compared to those without intellectual/developmental disabilities (51/135, 38% and median = 9 hours, respectively) (51/134, 38%) ($P=.038$). Parents and other caregivers of males with intellectual/developmental disabilities who are unable to adequately verbalize their testicular/groin/scrotal pain should be cognizant of the signs and symptoms associated with testicular torsion, perform a genitalia examination, and seek an immediate evaluation to diagnose and treat this urgent condition.

Keywords

pediatrics, developmental disability, intellectual disability, orchiectomy, urology, testicular torsion

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Introduction

Testicular torsion refers to twisting of blood vessels leading to the testicle resulting in lack of testicular blood flow.^{1,2} Accounting for 10% to 15% of acute scrotal disease in children, testicular torsion affects 1 in 4000 males <25 years of age.³⁻⁵ This condition represents a pediatric emergency as testicular pain lasting more than 4 to 8 hours is often associated with testicular death.^{1,5,6} Unilateral scrotal pain of sudden onset is the most common presenting symptom, and surgical intervention is the mainstay of treatment. Expediting surgery decreases the ischemic time and reduces the likelihood of testicular loss.^{7,8} Testicular salvage rates are 80% to 100% if the patient is surgically treated within 6 hours of symptom onset but plummet to 20% after 12 hours.^{3,4,8} Despite timely surgical intervention, approximately 33% to 44% of boys undergo an orchiectomy due to a nonviable testicle.^{1,3} Patients who have orchiectomies are often hospitalized for longer periods of time which results in

higher healthcare costs.⁹ Delayed and incorrect diagnosis are the primary reasons for unsalvageable testes.³

Prompt and accurate diagnosis of testicular torsion is imperative to avert testicular loss. Several factors are associated with delayed or inaccurate diagnosis of testicular torsion which increase the likelihood that the affected testicle will not be salvaged, including isolated abdominal pain, failure to perform a thorough genitourinary exam, scrotal imaging, late presentation to the Emergency Department (ED), and pediatrician evaluation prior to the ED.⁹⁻¹²

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A child is diagnosed with a developmental delay when he/she does not attain developmental milestones compared to peers of the same age range.¹³ Approximately 10% to 15% of children <5 years old will experience developmental delays that manifest in varying degrees of severity.¹³ Developmental disabilities refer to severe, lifelong impairment in development that interfere with learning, self-sufficient, and adaptive skills. Not all children with developmental delays will have developmental disabilities. As children with intellectual and developmental disabilities often have communication deficits,¹⁴ diagnosing and treating testicular torsion in a timely manner poses a challenge. To our knowledge, only one previous study addressed the connection between testicular torsion and developmental, cognitive, or social disorders.¹⁰

The present study describes several reasons that may contribute to the delay in diagnosis or misdiagnosis of testicular torsion, with a particular emphasis on patients with intellectual and developmental disabilities who have an inability to adequately verbalize their pain. Educating parents and other caregivers of male children and adolescents with intellectual and developmental disabilities about the importance of early detection of signs and symptoms of testicular torsion to expedite surgical intervention is discussed. As it is difficult to obtain specific information about the location of pain from children and adolescents with intellectual and developmental disabilities who are unable to verbalize their symptoms, we emphasize that parents and caregivers should be alert to behavioral changes, aware of the entity of testicular torsion, and evaluate the genitalia.

Materials and Methods

Under an Institutional Review Board-approved protocol and conforming to the World Medical Association Declaration of Helsinki, we identified male children and adolescents ages 1 to 18 years over a 6-year period (January 1, 2015–December 31, 2020) who experienced testicular torsion and had intellectual and developmental disabilities. The patients were identified by performing a medical record review of consecutive cases of testicular torsion seen at our Institution's ED. None of the patients was able to adequately verbalize testicular/groin/scrotal pain which led to a delay in diagnosis, longer duration of ischemia, and poorer outcome. The duration of symptoms was determined based on when the parent or other caregiver detected the abnormal symptoms.

All patients were evaluated at our Institution's ED by a pediatric urologist who obtained the medical history and performed a focused genitourinary physical examination. A pediatric urologist was available at all times to evaluate and treat patients with testicular

torsion. Four pediatric urologists are employed at our Institution, and the on-call schedule alternates between all 4. A Doppler scrotal US was performed on all patients. Numerous metrics were collected including the patient's age, duration and side of symptoms, testicular pain, scrotal swelling and erythema, absence of cremasteric reflex, high riding testicle, type of surgery (orchiectomy vs orchiopexy), and degree of testicular torsion. The patients' specific intellectual and developmental disabilities were also recorded. The parents of the patients with testicular torsion provided written informed consent.

Statistical Analysis

Continuous and categorical data were summarized by median (IQR) and count (%), respectively. Fisher's Exact test compared categorical characteristics between patients with intellectual and developmental disabilities and those without this diagnosis. Wilcoxon Rank Sum test similarly compared the continuous variables. *P*-values $\leq .05$ were considered statistically significant. All analyses were performed using R software (4.0.0).¹⁵

Ethical Approval and Informed Consent

The University of Louisville Institutional Review Board (IRB Number 20.0778) determined that our study was exempt according to 45 CFR 46.101(b) under Category 4. The patients' mothers provided written informed consent.

Results

Details of Intellectual and Developmental Delays in Patients With Testicular Torsion

Of the 140 patients between the ages of 1 to 18 years with testicular torsion over the 6-year period of this study, 5 (3.6%) had intellectual and developmental disabilities with an inability to verbalize pain. Table 1 depicts the specific intellectual and developmental disabilities in these 5 patients. All 5 patients had ≥ 1 intellectual and developmental disabilities. Case #3, an 18-year-old with cerebral palsy and quadriplegia, was undergoing a diaper change 1 night prior to his ED admission at which time his mother detected left groin swelling and her son's discomfort with palpation.

Characteristics of Patients With Testicular Torsion Who Have Intellectual and Developmental Disabilities

The median age of patients with intellectual and developmental disabilities was 14.0 years (range 13–16 years).

Table 1. Details of Intellectual and Developmental Delays in Patients With Testicular Torsion at Our Institution (January 1, 2015-December 31, 2020).

Case #	Age (years)	Duration of symptoms (hours)	Presenting symptoms	Type of surgery	Degree of testicular torsion	Intellectual and/or developmental delays
1	16	96	96 hours: left testicular swelling and pain, dysuria, walk with wide-based gait 48 hours: dysuria 6 hours: testicular pain	Left orchiectomy	540	ADHD, autism spectrum disorders, intellectual disabilities
2	13	72	Abdominal discomfort, right scrotal swelling, right groin pain	Right orchiectomy	360	ADHD, chromosome abnormalities, developmental disorder, lack of expected normal physiological development, intellectual disabilities, mixed receptive-expressive language disorder
3	18	12	Left groin swelling	Left orchiectomy	1080	Cerebral palsy, quadriplegic, developmental delay, grinding of teeth
4	14	24	Right testicular pain and swelling	Right orchiectomy	360	ADHD, autism spectrum disorder, speech delay
5	13	20	Left testicular swelling, pain, erythema	Left orchiectomy	360	Spastic cerebral palsy, developmental delay, epilepsy, hydrocephalus

Abbreviation: ADHD, attention deficit hyperactivity disorder.

The patients with intellectual and developmental disabilities (5/5, 100%) underwent significantly more orchiectomies compared to those without intellectual/developmental disabilities (51/135, 38%) ($P=.009$) (Table 2). Additionally, the median duration of symptoms was significantly higher for patients with intellectual and developmental delays compared to those without intellectual/developmental disabilities (48 vs 9 hours, respectively) ($P=.047$). Of the 5 patients with intellectual and developmental delays, the shortest duration of symptoms was 12 hours, and the longest was 96 hours. Significantly more patients with intellectual and developmental disabilities (5/5, 100%) had a symptom duration of ≥ 12 hours when compared to those without intellectual/developmental disabilities (91/135, 67%) ($P=.018$).

The most common symptoms were testicular/groin/scrotal swelling in all 5 patients and testicular/groin pain in 4 patients. The degrees of testicular torsion ranged between 1 and 3 full circles (360°-1080°).

Discussion

The most common causes of testicular loss after torsion are reportedly due to delays in seeking medical attention

(58%), incorrect initial diagnosis (29%), and treatment delays at a referral hospital (13%).¹⁶ Several elements have been associated with inaccurate or delayed diagnosis of testicular torsion, including late presentation to the ED, pediatric evaluation before being seen in the ED, scrotal imaging, isolated abdominal pain, and failure to perform a complete genitourinary examination.^{10,11}

As the most frequent presenting symptoms of testicular torsion are unilateral testicular pain, nausea, and vomiting, providers may be less likely to link isolated abdominal pain to testicular torsion. In Bayne et al¹⁰ study of 208 patients who underwent surgery for testicular torsion, 114 (54.8%) patients presented in the acute setting (<24 hours), while 94 (45.2%) (>24 hours) were delayed. A total of 19 (20.2%) patients in the delayed group reported isolated abdominal pain and no scrotal pain, whereas no patients in the acute group had only isolated abdominal pain ($P<.001$). Failure to perform a genitourinary exam was associated with misdiagnosis. These authors recommend a thorough genitourinary examination in all patients with isolated abdominal pain and that the provider should consider testicular torsion in the differential diagnosis for these patients.

Scrotal imaging is the gold standard for diagnosing testicular torsion due to its non-invasive and accessible

Table 2. Characteristics of Patients With Testicular Torsion Who Have Intellectual and Developmental Disabilities at Our Institution (January 1, 2015-December 31, 2020).

Characteristic	Intellectual and developmental disabilities		P-value
	No N = 135	Yes N = 5	
Age (years)	13.0 (12.0, 15.0)	14.0 (13.0, 16.0)	.204
Duration of symptoms (hours)	9.0 (5.0, 48.0)	48.0 (20.0, 72.0)	.047
Duration of symptoms \geq 6 hours	91 (67%)	5 (100%)	.325
Duration of symptoms \geq 12 hours	59 (44%)	5 (100%)	.018
Duration of symptoms \geq 24 hours	42 (31%)	3 (60%)	.328
Testicular pain = Yes	132 (98%)	4 (80%)	.137
Scrotal swelling = Yes	91 (69%)	5 (100%)	.322
Scrotal erythema = Yes	42 (32%)	2 (40%)	.658
Absent cremasteric reflex = Yes	42 (31%)	0 (0%)	.322
High riding testicle = Yes	41 (31%)	2 (40%)	.651
Side of torsed testis			1.000
Both	2 (1%)	0 (0%)	
Left	75 (56%)	3 (60%)	
Right	58 (43%)	2 (40%)	
Surgery = orchiectomy	51 (38%)	5 (100%)	.009
Degrees of torsion	360 (315, 720)	360 (360, 540)	.671

The bold values in table are the *P*-values that are statistically significant.

nature, lack of radiation exposure, and high sensitivity (98%-100%) and moderate to high specificity (69%-98%).^{3,10} Confirmatory findings demonstrate decreased or lack of blood flow to the affected testicle. While it has been reported that scrotal imaging may contribute to the delay in surgically treating testicular torsion and result in increased orchiectomy rates,^{3,9} failure to perform this test often leads to misdiagnosis of this urgent condition.¹⁰

To our knowledge, only one previous study described delayed diagnosis or misdiagnosis of testicular torsion in patients with developmental, cognitive, or social disorders. In Bayne et al¹⁰ study, patients in the delayed presentation group were 4 times more likely to have a developmental, cognitive, or social disorder than those in the acute presentation group (10.6% vs 2.6%; *P* = .02). One-half of the patients in the delayed setting group were diagnosed with autism spectrum disorder. Twelve patients in the delayed presentation group were initially misdiagnosed, all of whom eventually underwent an orchiectomy. Developmental, cognitive, or social disorders were more frequent in the misdiagnosed group than the acute group (25% vs 2.6%; *P* = .02). These authors attributed these patients' inability to clearly communicate about their symptoms for both the delayed diagnosis and misdiagnosis of testicular torsion.

Similar to patients who have intellectual and developmental delays, younger children with testicular torsion

may also have difficulty communicating their symptoms to their caregivers. In Ramachandra et al¹⁷ study of 114 patients with testicular torsion, patients older than 8 years of age were more likely to undergo orchidopexy than those younger than 8 (61.5% vs 30.4%, *P* = .01). Younger children may not have noticeable physical findings such as scrotal swelling or firmness until later in the disease process which may increase the likelihood that the testis will not be salvageable. Furthermore, as young children are unable to effectively articulate their symptoms, the duration of symptoms may be longer than that reported by their caregivers.

Strengths and Limitations

Our 6-year study of testicular torsion highlighting patients with intellectual and developmental disabilities emphasizes the importance of recognizing the symptoms associated with testicular torsion in this unique population and expediting the diagnosis and treatment of this pressing condition. There were significantly more orchiectomies and a significantly longer duration of symptoms in patients with intellectual and developmental disabilities compared to those without intellectual/developmental disabilities. Our study serves as a valuable instructional message to parents and caregivers of male children and adolescents with intellectual and developmental disabilities to be familiar with the

symptoms associated with testicular torsion such as scrotal/testicular pain and swelling and be on heightened alert for signs of pain/discomfort as possible presenting symptoms of testicular torsion in patients who are non-verbal. Additionally, parents and caregivers should be vigilant of ongoing unusual behaviors that may indicate testicular torsion. For example, the symptoms suggestive of testicular torsion in Case #1 persisted for several days prior to ED evaluation: the patient walked with a wide-based gait 96 hours prior, experienced dysuria 48 hours prior, and complained of testicular pain 6 hours prior. As this patient suffered from autism spectrum disorder and intellectual disabilities, he may have been unable to convey the extent and severity of his symptoms in an expeditious manner which subsequently led to a required orchiectomy.

A thorough history and scrotal US are necessary in patients with intellectual and developmental disabilities to confirm testicular torsion. Since all 5 of the patients had testicular/groin/scrotal swelling and 4 had testicular/groin pain in our study, it is also crucial that caregivers evaluate the genitalia. If testicular torsion is a concern, the child/adolescent should be evaluated promptly in the ED.

The limitations of the current study are its retrospective nature and limited number of patients with intellectual and developmental disabilities who developed testicular torsion. Additional evaluations of a larger number of patients with intellectual and developmental disabilities who experience testicular torsion are warranted to further investigate how these patients may be diagnosed and treated expeditiously.

Conclusion

Restoring testicular blood flow in a timely manner is the ultimate goal in salvaging the affected testis in patients who develop testicular torsion. Male children and adolescents with intellectual and developmental disabilities are often unable to effectively express their symptoms of testicular torsion which significantly delays diagnosis and rapid surgical intervention. Parents and other caregivers of children with intellectual and developmental disabilities who are deficient in verbal ability to convey testicular/groin/scrotal pain should be aware of any unusual signs of pain/discomfort/behavioral changes, be educated about the signs and symptoms of testicular torsion, and pursue prompt evaluation to reduce the likelihood of an orchiectomy.

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Author Contributions

LBES: Data conception, design, acquisition, analysis, and interpretation; drafted the manuscript; critically revised the manuscript and gave final approval.

MWD: Data conception, design, acquisition, analysis, and interpretation; critically revised the manuscript and gave final approval.

DSP: Data conception, design, acquisition, analysis, and interpretation; critically revised the manuscript and gave final approval.

ER: Data conception, design, acquisition, analysis, and interpretation; critically revised the manuscript and gave final approval.

Declaration of Conflicting Interests

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Informed Consent

The patients' mothers provided written informed consent.

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References

1. DaJusta DG, Granberg CF, Villanueva C, Baker LA. Contemporary review of testicular torsion: new concepts, emerging technologies and potential therapeutics. *J Pediatr Urol*. 2013;9:723-730.
2. Shields LBE, Daniels MW, Peppas DS, et al. Surge in testicular torsion in pediatric patients during the COVID-19 pandemic. *J Pediatr Surg*. Published online July 16, 2021. doi:10.1016/j.jpedsurg.2021.07.008
3. Chan EP, Wang PZ, Myslik F, Chen H, Dave S. Identifying systems delays in assessment, diagnosis, and operative management for testicular torsion in a single-payer health-care system. *J Pediatr Urol*. 2019;15:251.e1-251.e7.
4. Ringdahl E, Teague L. Testicular torsion. *Am Fam Physician*. 2006;74:1739-1743.
5. Sharp VJ, Kieran K, Arlen AM. Testicular torsion: diagnosis, evaluation, and management. *Am Fam Physician*. 2013;88:835-840.
6. Osumah TS, Jimbo M, Granberg CF, Gargollo PC. Frontiers in pediatric testicular torsion: an integrated review of prevailing trends and management outcomes. *J Pediatr Urol*. 2018;14:394-401.
7. Arevalo MK, Sheth KR, Menon VS, et al. Straight to the operating room: an emergent surgery track for acute testicular torsion transfers. *J Pediatr*. 2018;192:178-183.

8. Morin OA, Carr MG, Holcombe JM, Bhattacharya SD. Optimal predictor of gonadal viability in testicular torsion: time to treat versus duration of symptoms. *J Surg Res.* 2019;244:574-578.
9. Steeman A, Ngatchou W, Ramadan AS, et al. Impact of treatment delays on outcome of acute testicular torsion: a 15-year retrospective study. *Acta Chir Belg.* Published online February 14, 2021. doi:10.1080/00015458.2021.1883391
10. Bayne CE, Villanueva J, Davis TD, Pohl HG, Rushton HG. Factors associated with delayed presentation and misdiagnosis of testicular torsion: a case-control study. *J Pediatr.* 2017;186:200-204.
11. Rampaul MS, Hosking SW. Testicular torsion: most delay occurs outside hospital. *Ann R Coll Surg Engl.* 1998;80:169-172.
12. Ryan KA, Folkard SS, Bastianpillai C, Green JSA. The management of testicular torsion in the UK: how can we do better? A national quantitative and qualitative analysis of the factors affecting successful testicular salvage. *J Pediatr Urol.* 2020;16:815.e1-815.e8.
13. Choo YY, Agarwal P, How CH, Yeleswarapu SP. Developmental delay: identification and management at primary care level. *Singapore Med J.* 2019;60:119-123.
14. Rogers SJ. Diagnosis of autism before age three. In: Glidden L (ed.) *International Review of Research in Mental Retardation: Autism.* 23rd ed. Academic Press; 2000:1-31.
15. R Core Team. R: a language and environment for statistical computing. R Foundation for Statistical Computing. Accessed September 22, 2021. <https://www.R-project.org>
16. Matteson JR, Stock JA, Hanna MK, Arnold TV, Nagler HM. Medicolegal aspects of testicular torsion. *Urology.* 2001;57:783-786; discussion 786-787.
17. Ramachandra P, Palazzi K, Holmes N, Marietti S. Factors influencing rate of testicular salvage in acute testicular torsion at a tertiary pediatric center. *West J Emerg Med.* 2015;16:190-194.