

## Perinatal Testicular Torsion with Bilateral Radioulnar Synostosis: Case Report

Mohammed Al-Biltagi<sup>1\*</sup>, Fayza Haider<sup>2</sup> and Fatima Naser AbdulAziz<sup>3</sup>

<sup>1</sup>Professor of Pediatrics, Pediatric Department, Tanta University, Egypt

<sup>2</sup>Consultant Pediatric Surgeon, Salmaniya Medical Complex, Ministry of Health, Kingdom of Bahrain

<sup>3</sup>Consultant Orthopedic Surgeon, Salmaniya Medical Complex, Ministry of Health, Kingdom of Bahrain

**\*Corresponding Author:** Mohammed Al-Biltagi, MD, PhD, Pediatric Department, Faculty of Medicine, Tanta University, Qism 2, Tanta 11432, Egypt.

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### Abstract

Perinatal testicular torsion and congenital bilateral radioulnar synostosis are rare neonatal disorders. Four days-old male neonate of diabetic mother presented with painless left testicular swelling and restricted mobility of both elbows and wrist. Scrotal ultrasound revealed enlarged left testicle with small hypo echogenic necrotic areas and absent perfusion of the spermatic vessels on the left side by Doppler ultrasound. Plain film radiography showed congenital bilateral radioulnar synostosis. He underwent scrotal exploration, immediate orchidectomy with contralateral orchidopexy. Conclusions: Perinatal testicular torsion and congenital radioulnar synostosis are rare neonatal disorders. Their association together is never described before. Proper orientation of the pediatricians and high levels of suspicion will help to detect these disorders as early as possible.

**Keywords:** Perinatal Testicular Torsion; Congenital Bilateral Radioulnar Synostosis

### Core tip

Perinatal testicular torsion and congenital bilateral radioulnar synostosis are rare neonatal disorders. It is the first time to describe this association. Awareness of the pediatricians with these disorders will help their early recognition and proper management especially for the testicular torsion.

### Introduction

Both testicles and bones develop from the mesoderm. The testicles develop from the meso-dermal bud proximal to the kidney [1], while radius and ulna develop from the mesodermal upper limb bud at 35 - 37 days after fertilization [2]. Torsion of the spermatic cord in neonates (perinatal torsion) is very rare, and occurs in 1:7500 newborn. It is recently subcategorized as either occurring prenatally in utero or postnatally in the first month of life [3]. Radioulnar synostosis arises due to longitudinal segmentation failure and persistence of the cartilaginous primitives between the radius and ulna at 35 to 37 days of development resulting in persistent tissue bridge between the two bones as an unsegmented block, usually at the proximal level [4]. According to our best knowledge, this is the first case reported association of perinatal testicular torsion with congenital radioulnar synostosis.

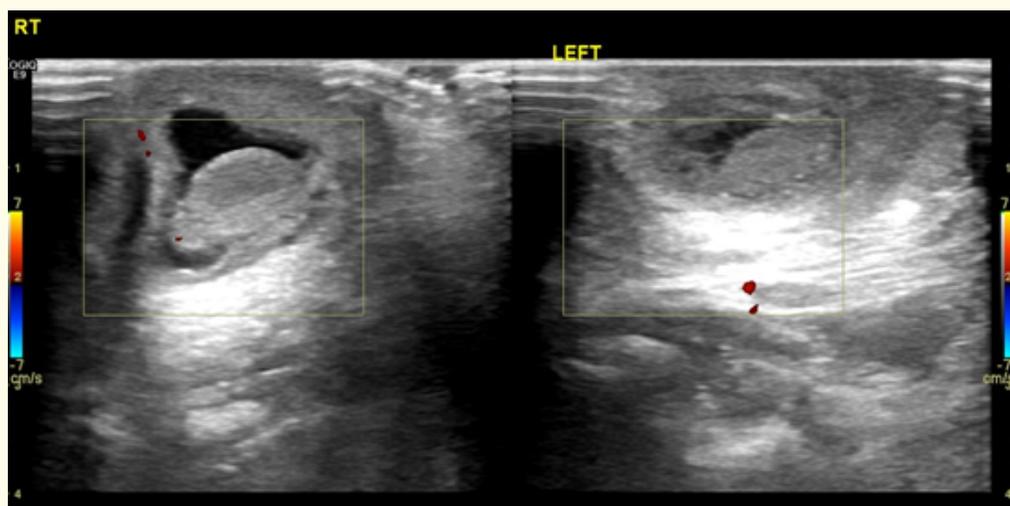
### Case Presentation

A four-day old male neonate presented to us for evaluation of neonatal jaundice. During examination, we found a painless firm mass on his left scrotum. He was born by elective lower segment caesarean section (LSCS) at 38 weeks of gestation due to previous three CS. His birth weight was 3.652 kg and his APGAR score was 8 at 1 minutes and 10 at 5 minutes. He is the product of non-consanguineous marriage

of 39 yrs old mother and 57 years old father. The mother developed gestational diabetes controlled by both diet and Insulin therapy and hypertension controlled by salt restriction and Methyldopa.

On examination, the neonate had mild hypotonia, jaundice, and excessive tearing from both eyes. He kept his both forearm in pronation, and it was not possible to do passive supination, with no rotation of forearm. However, the wrist, elbow neck, hip, knee, and feet motion, were normal. The left hemiscrotum was swollen with erythematous, bluish tinge. The left testis was higher in position related to its upper pole and showed painless, firm, non- fluctuating swelling that did not transilluminate. There was right mild hydrocele. Cremastic reflex was absent on the left side but present on the right side. The spermatic cords were not thickened.

Scrotal ultrasound (Figure 1) revealed enlarged left testicle with small hypo echogenic necrotic areas inside the left testis together with hyper echoic area surrounding the upper side of the left scrotum at the level of the left epididymis suggesting organized hematoma. Doppler ultra sound did not detect any perfusion of the spermatic vessels on the left side. There was mild free fluid seen in the right scrotum suggesting a hydrocele measuring approximately 5 mm in maximum dimension. The right testis and epididymis were normal in echogenicity, size, outline and vascularity. No hypervascularity was appreciated that may suggest active inflammation. Power Doppler imaging of the testicles, however, revealed absent central flow in the left testis with only peripheral flow suggested non-viability of the left testicle. Both kidneys were normal without organomegaly. X-ray of forearms (Figure 2) showed fusion of the proximal portion of the ulna and radius and ulna on both sides suggestive of bilateral total synostosis present between the proximal end of the radius and ulna without subluxation or dislocation of the elbow joints (Figure 2). Immediate scrotal exploration showed unsalvageable left testis so; left orchidectomy and right orchidopexy were done. The orthopedic surgeon suggested follow up of his skeletal deformity for correction before school age.



**Figure 1:** Scrotal Doppler Ultrasound.

*It showed hyper echoic to content is seen surrounding the upper side of the left scrotum at the level of the left epididymis suggesting organized hematoma (acute to sub-acute) with small hypoechogenic areas seen in the left testis. Doppler sound could not detect any perfusion of the spermatic vessels on the left side. There was mild free fluid seen in the right testis suggesting a hydrocele measuring approximately 5 mm in maximum dimension. The right testis and epididymis were normal in echogenicity, size, outline and vascularity.*



**Figure 2:** Forearm AP/LAT.

*It showed synostosis between the proximal end of the radius and ulna.*

### Discussion

We described a male newborn with unilateral perinatal testicular torsion and bilateral radioulnar synostosis, with typical clinical signs, ultrasound features, and X-ray finding. It is the first time to find this association according to our literature review on all the available MEDLINE search data sources. In this case, the torsion presented as painless swelling with dusky erythematous, bluish discoloration of the scrotal skin which indicate prenatal onset of the torsion, unlike the postnatal torsion which usually present with acute manifestation with considerable tenderness and swelling of a previously normal testicle [5].

High birth weight (60% are above the 90<sup>th</sup> centile for weight), gestational diabetes, multi-parity, over reactive cremasteric reflex, difficult labor and breech presentation are risk factors for perinatal testicular torsion [6]. In the present case, the infant had birth weight of 3.652 kg and was the product of multiparous mother with gestational diabetes and hypertension. It is of paramount importance to recognize the torsion early and to differentiate between prenatal and postnatal onset testicular torsion, as the management is quite different. Most cases of perinatal testicular torsion appear prenatally, and testicles are unsalvageable [6] while the postnatal testicular torsion is an acute state and needs immediate surgery. In many cases, it is difficult to differentiate between the two types of torsions especially with delayed diagnosis. Therefore, immediate exploration and surgery is a standard procedure in acute perinatal testicular torsion; despite presence of extensive arguments concerning management of the “old” prenatal testicular torsions; from immediate orchiectomy to conservative treatment resulting in testicle atrophy [7]. Table 1 showed comparison between prenatal and postnatal onset types of testicular torsion.

	Prenatal (72%)	Postnatal (28%)
A/E	- Difficult labor	- Breech presentation
	- High birth weight (60% are above the 90 <sup>th</sup> centile for weight)	
	- Over reactive cremasteric reflex	- Gestational Diabetes
	- Multi-parity	
Laterality	Almost always Unilateral	Usually Unilateral
Type	Extra-vaginal	Extra-vaginal
Onset	In utero & appears since birth	After birth in the first month of life
Symptoms	Minimal pain to no discomfort	Acute manifestation severe pain, nausea & vomiting /acute abdomen
Signs	Few localized findings e.g. swelling, discoloration, no erythema/oedema. Testicular agenesis ('vanishing testis')	considerable tenderness & redness with swelling of a previously normal testicle
Fate	Testis is almost always necrotic Testicular agenesis	The testicles can be salvaged if immediate surgery is done

**Table 1:** Comparison between prenatal onset and postnatal onset testicular torsion.

Immediate surgery has the advantages of being able to confirm torsion and to distinguish it from other causes of acute scrotal pain and swelling and the ability to perform contralateral orchidopexy to decrease the likelihood of a contralateral extravaginal torsion during the first 2 months of life. Despite being safe, non-lengthy procedure with minimal intraoperative risks, it still carries higher chance of injury to the single testis [8]. On the other hand, some researches may prefer conservative treatment as some testicular tissue (especially the Leydig cells) can survive after torsion, even if perfusion does not resume. In addition, conservative treatment avoid various surgical and anesthetic-related risks in the neonatal period. However, it carries the risk of possible damage of contralateral healthy testicle through antisperm antibody production [9]. Table 2 showed comparison between surgical and conservative management of perinatal onset testicular torsion.

	Immediate surgery	Conservative TTT
Indication	Prenatal/Acute Standard Procedure	Old torsion (very low occurrence of contralateral torsion)
Results	Surgical removal	testicle atrophy
Advantages	<ul style="list-style-type: none"> <li>- Confirm the torsion</li> <li>- Ruling out other possible diagnoses.</li> <li>- Decrease the likelihood of a contralateral extravaginal torsion during the first 2 months of life.</li> <li>- Safe, non-lengthy procedure</li> <li>- Minimal intraoperative risks</li> </ul>	<ul style="list-style-type: none"> <li>- Some tissue after torsion can survive, even if perfusion does not resume - this concerns especially the Leydig cells.</li> <li>- Avoid various surgical and anesthetic-related risks in the neonatal period</li> </ul>
Disadvantages	-Higher chance of injury to the single testis	- The possibility of contralateral healthy testicle damage through antisperm antibody production

**Table 2:** Comparison between surgical and conservative management of perinatal onset testicular torsion.

Radiaoulnar synostosis is usually congenital; but it may also occur after a forearm fracture or trauma. Congenital proximal radiaoulnar synostosis is an uncommon malformation, often bilateral (52.9%), and more commonly seen in male patients (64.7%). In congenital radiaoulnar synostosis there is fusion of the proximal ends of the radius and ulna, fixing the forearm in varying degrees of pronation [10]. In the presenting case, the fusion was about 3 cm at the proximal end due to error during early limb development. The limb bud will form humerus, radius, and ulna. Certain hormonal or timing factors may not signal the radius and ulna to form into two separate and discrete bones. If the bones do not properly form, the muscles that rotate the radius and ulna are frequently absent. With growth and development, the cartilaginous union ossified and becomes a bony bridge, between the radius and ulna and continue to share a common perichondrium. The synchondrosis usually ossifies between 1 and 4 years old [11,12]. This abnormality may occur as an isolated disorder or associated with other skeletal (in one third of cases) or associated with systemic problems (cardiac, renal, neurologic or gastrointestinal disorders). It is also commonly associated with certain genetic syndromes, such as Holt-Oram syndrome (hand-heart syndrome) and fetal alcohol syndrome. A genetic factor is incriminated in 25% of patients [13,14].

This is the first report to describe an association of perinatal testicular torsion with congenital bilateral proximal radiaoulnar synostosis. Common etiological factors may be the reason of this association. Gestational diabetes is a common etiological factor in both conditions [5,15]. Multiparity is predisposing for perinatal testicular torsion [5] while old maternal age is predisposing for congenital radiaoulnar synostosis [15]. In our case both multiparity and old maternal age are present.

Unfortunately, chromosomal study was not done in the presented case. It is important to rule out chromosomal abnormality that could be associated with perinatal testicular torsion or radiaoulnar synostosis. The testis-determining SRY region present on chromosome Y directly affects the differentiation of the indifferent gonad into a testis shortly after 6 weeks' gestation. Testicular torsion was reported to be an autosomal or X-linked recessive mode of inheritance for at least some of the instances of supravaginal torsion of the spermatic cord [16]. At the same time, radiaoulnar synostosis was also reported in cases with X-chromosome disorders such as Klinefelter syndrome. Sex chromosome polysomy is associated with triad of mental retardation, skeletal malformation, and hypogonadism [17]. In the presence of maternal gestational diabetes, there is an increased risk for chromosomal aneuploidy as well as for congenital anomalies in the offspring. Screening of other congenital anomalies should be done to rule out any associated cardiac, renal, and neurologic anomalies. Serial follow up of bone marrow function is mandatory to detect any potential bone marrow hypoplasia as early as possible.

## Conclusion

Despite perinatal testicular torsion and congenital radioulnar synostosis are neonatal disorders, the pediatrician should be aware of their presentation. Higher level of suspicion may help to decrease the rate of missing these rare abnormalities. Early surgical intervention may save one or both testicles.

**Author contributions:** Al-Biltagi M, Haider F, and AbdulAziz F designed the report; Haider F performed the surgery; AbdulAziz F did the orthopedic diagnosis and Al-Biltagi M collected the patient's clinical data; and wrote the paper. All the three authors revised the manuscript.

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## Bibliography

1. Archambeault DR., *et al.* "Epithelial-mesenchymal crosstalk in Wolffian duct and fetal testis cord development". *Genesis* 47.1 (2009): 40-48.
2. Shinohara T., *et al.* "Painful snapping elbow in patients with congenital radioulnar synostosis: report of two cases". *Journal of Hand Surgery American* 35.8 (2010): 1336-1339.
3. Guerra LA., *et al.* "Management of neonatal testicular torsion: Which way to turn?". *Canadian Urological Association Journal* 2.4 (2008): 376-379.
4. CR Bhatt and CD Mehta. "Case Report: Congenital Radioulnar Synostosis and Its Embryological Correlation and Functional Assessment". *Journal of Anatomical Society of India* 60.2 (2011): 236-238.
5. Muhammad Riaz-Ul-haq., *et al.* "Neonatal Testicular Torsion; a Review Article". *Iranian Journal of Pediatrics* 22.3 (2012): 281-289.
6. Piet R H Callewaert and Philip Van Kerrebroeck. "New insights into perinatal testicular torsion". *European Journal of Pediatrics* 169.6 (2010): 705-712.
7. Djahangirian O., *et al.* "Timing and surgical management of neonatal testicular torsions". *Journal of Pediatric Surgery* 45.5 (2010): 1012-1015.
8. Kaye JD., *et al.* "Neonatal torsion: a 14-year experience and proposed algorithm for management". *Journal of Urology* 179.6 (2008): 2377-2383.
9. Rybkiewicz M. "Long-term and late results of treatment in patients with a history of testicular torsion". *Annales Academiae Medicae Stetinensis* 47 (2001): 61-75.
10. Hung NN. "Derotational osteotomy of the proximal radius and the distal ulna for congenital radioulnar synostosis". *Journal of Children's Orthopaedics* 2.6 (2008): 481-489.
11. Boulet AM and Capecchi MR. "Multiple roles of Hoxa11 and Hoxd11 in the formation of the mammalian forelimb zeugopod". *Development* 131.2 (2004): 299-309.

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12. Elliott AM., *et al.* "The developmental spectrum of proximal radioulnar synostosis". *Skeletal Radiology* 39.1 (2010): 49-54.
13. Siemianowicz A., *et al.* "Congenital radioulnar synostosis - case report". *Polish Journal of Radiology* 75.4 (2010): 51-54.
14. Yammine K., *et al.* "Congenital radioulnar synostosis. Study of a series of 37 children and adolescents". *Chirurgie de la Main* 17.4 (1998): 300-308.
15. Castillo-Caro P., *et al.* "Proximal radio-ulnar synostosis with bone marrow failure syndrome in an infant without a HOXA11 mutation". *Journal of Pediatric Hematology/Oncology* 32.6 (2010): 479-485.
16. Castilla EE., *et al.* "Neonatal testicular torsion in two brothers". *Journal of Medical Genetics* 12.1 (1975): 112-113.
17. Sumathipala D., *et al.* "An infant born to a mother with gestational diabetes presenting with 49, XXXXY syndrome and renal agenesis-a case report". *Journal of Clinical Research in Pediatric Endocrinology* 4.4 (2012): 223-225.

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