

Congenital gluteus maximus contracture syndrome - a case report with review of imaging findings

Vamshi Krishna Kotha^{1*}, Rajasekhar Reddy¹, M. Venkateshwar Reddy², (Brig) Rangubatla Sathyanarayana Moorthy¹, Tatikonda Venkat Kishan¹

1. Department of Radiology, Kamineni Institute of Medical Sciences, Andhra Pradesh, India

2. Department of Orthopaedics, Kamineni Institute of Medical Sciences, Andhra Pradesh, India

* Correspondence: Dr. Kotha Vamshi Krishna, MD, #1504, 411 Duplex Avenue, Toronto, ON M4R 1V2, Canada
(✉ vamshikotha@gmail.com)

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ABSTRACT

Although the clinical features of gluteus maximus contracture syndrome have been frequently described, imaging features have been seldom described. Most commonly reported cases are those following intramuscular injection in the gluteal region although congenital contracture is an uncommon but important occurrence. This condition has most often been reported in children of school going age. These patients often present with difficulty in squatting, limitation of hip motion or specific deformities and often require surgical correction. We describe the plain radiography, ultrasonography (USG) and magnetic resonance imaging (MRI) features of this condition in a patient with no previous known history of intramuscular injections.

CASE REPORT

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Presentation

A 9 year old male child presented to us with a history of difficulty in squatting and an abnormal 'legs-apart' sitting posture with a gradually increasing waddling gait since childhood. He was born from a non-consanguineous marriage by normal vaginal delivery. The parents gave no history of repeated intramuscular injections other than vaccines received in the neonatal period. There was no history of any other illness in the past or any previous hospital admission.

Clinical Features

On examination, the child was of normal height and build for age. Abnormal squatting posture was noted with both hips in an abducted, externally rotated position (Figure 1A). He was unable to approximate his knees in squatting position without losing balance. There was restriction of flexion of both hips, left more severe than right, along with weakness of the abductors on both sides. Flattening of the buttocks was also observed (Figure 1B). A clinical diagnosis of bilateral hip abductor abnormality was made and the child was referred for radiological investigations.

Imaging features

An anteroposterior radiograph of the pelvis (Siemens 600 mA, 80 kV, 20 mAs) revealed a subtle curved line along the lateral iliac cortex adjacent to the left sacroiliac joint (Figure 3A and 3B). Ultrasonography (Siemens Acuson X300, 8 MHz, linear transducer) revealed decrease in the muscle bulk of both glutei maximus (left worse than right) with increase in echorefectivity of the upper and middle 1/3rd of the left gluteus maximus muscle suggestive of fibrosis (Figure 3D).

MRI (Siemens Essenza, 1.5T) scan was performed with a standard body matrix and patient in a supine position (Figure 2). The following pulse sequences were obtained: T1 spin echo at TR/TE 772/12, 3 mm slice thickness, FOV 36 cm, Matrix 384 x 280; T1 thin section MPR at TR/TE 1800/12, 1 mm slice thickness, FOV 36 cm, Matrix 384 x 280; T2 spin echo at TR/TE 1500/127, 1 mm slice thickness, FOV 36 cm, Matrix 320 x 256. There was marked atrophy of both the glutei maximus [left more than right] with presence of a thick low intensity band within the upper and middle 1/3rd of the belly of left gluteus maximus. There was medial retraction of the tendons of both the involved muscles at their attachment sites

along with that of the adjacent iliotibial tracts. The gluteus medius and minimus on both sides were normal. A diagnosis of bilateral gluteus maximus contracture syndrome was made, most probably congenital in etiology.

Treatment

The patient underwent surgical release of bilateral gluteus maximus contractures and physiotherapy was suggested. Operative findings corroborated with the radiological findings and a thick contracture band was seen (Figure 4B). On follow up after 6 weeks of surgery, there was reduction in the abduction-external rotation deformity of the hips on squatting and the squatting and sitting actions had improved to a great extent (Figure 1C, 1D). Physiotherapy was continued for further improvement in hip function. Histopathological examination of the biopsied tissue revealed muscle atrophy and presence of fibrotic tissue within the muscle belly (Figure 4A).

DISCUSSION

Demographics

Gluteus maximus contracture syndrome is a debilitating disorder, often causing marked reduction in the quality of life. This condition most often occurs in children aged 6-18 years and is more common in boys than in girls [1,2]. A childhood incidence of 1-2.5% [3] and a morbidity of 1.36% in affected individuals has been reported [4]

Etiology

The most common etiological factor is multiple intramuscular injections into the gluteus maximus which may lead to muscular degeneration and fibrosis [5,6]. Ko et al reported a statistically significant association between intramuscular injections and gluteal fibrotic contractures in their case control study [7]. A similar condition has also been reported to occur in the form of contracture of the quadriceps femoris, deltoid and triceps brachii muscles which are all common sites of intramuscular injection [8,9]. Benzoyl alcohol used as a dissolvent in Penicillin has been reported to be an inciting factor [10,11,12].

A congenital variant of gluteus maximus contracture syndrome has also been described, similar to congenital torticollis. However, the pathogenesis and inheritance is poorly understood although presence of this syndrome in both parent and child has been reported in literature, substantiating the possibility of a congenital etiology [13].

Relevant Anatomy and Pathology

The gluteus maximus is a large, coarsely fasciculated muscle arising from the gluteal surface of the ilium, the gluteal aponeurosis, the sacroiliac ligament, the dorsal aspect of the sacrum and the upper portion of the coccyx. About 3/4ths of the muscular fibers coalesce to form a thick tendon that inserts onto the iliotibial tract and about 1/4ths of the fibers insert onto the gluteal tuberosity of the proximal femur [14].

In gluteus maximus contracture syndrome, the muscle undergoes degeneration and necrosis and is gradually replaced

by fibrosis causing restriction of contractility. On histopathological examination, the affected muscle shows fibrotic changes with collagen fibers replacing skeletal muscle fibers [15].

Clinical features

Fibrosis of the gluteus maximus leads to typical hip deformity which goes into abduction and external rotation on squatting and the knees cannot be approximated in this position when both hips are affected. There is flattening of the buttock on the affected side producing a 'cone shaped' deformity of the buttocks with bilateral hip affliction [3]. There is restriction of hip flexion with inability to cross the affected limb and difficulty in moving the knee towards the opposite shoulder.

Two clinical tests have been described - the squatting test and the active flexion test [16]. In the squatting test, the patient is asked to squat and then approximate his knees which he fails to perform due to bilateral hip affliction. In the flexion test, the patient is asked to flex only one hip while holding the other in an extended position. The test is positive if flexion is limited to less than 90o (hip flexion is usually 121+/-13o in healthy adults).

Zhao et al classified the syndrome into three levels based on the abduction contracture angles and three classes based on the involvement of the gluteus medius and minimus in addition to the maximus [13]. Another classification system was proposed by Ye et al which categorized the condition into four groups based on the clinical features which could predict the anatomic site of contracture and aid in planning of surgical correction [17]. Uncorrected contracture of the gluteus maximus can lead to serious deformities and functional disability in children including pelvic tilt, coxa valga and leg length discrepancy [18].

Imaging Findings

Plain radiographs do not show any significant changes early in disease progression. However, in advanced cases, an 'iliac hyperdense line' has also been described on pelvic AP radiographs and is said to be due to deformity of the lateral iliac cortex at the site of attachment of the gluteus maximus due to longstanding tugging effect of the contracted muscle [19]. The lateral cortex of the posterior ilium which is normally oriented oblique to the X-ray beam in an AP film is deformed into an orientation tangential to the X-ray beam and is then seen on the film as a vertical dense line. The subtle vertical curved line seen along the left lateral iliac cortex on the pelvic radiograph in our patient was thought to represent the vertical dense line described above.

Ultrasonographic features of this condition have also been reported including thinning of the involved muscle relative to that of a normal individual and presence of echogenic strips within the muscle bundles representing fibrous bands. The sensitivity of muscle thinning (92.6%) was higher than that of presence of echogenic strips (88.9%) but the former had a lower specificity (50%) than that of the latter (83.3%) [20]. Thinning of the gluteus maximus was seen on both sides in our

patient whereas an echogenic fibrous strip was demonstrated within the left gluteus maximus.

MRI is the modality of choice to delineate the extent of involvement of the affected muscles. Chen et al described various findings on MR imaging in this condition and classified them into primary and secondary features [16]. Similar to sonographic features, the primary MRI findings include atrophy of the gluteus maximus and presence of an intramuscular fibrous cord.

Muscle atrophy appears as thinning of the muscle belly with reduced compactness of the fiber bundles interspersed with fat seen as vertical hyperintense stripes on T1 and T2 weighted images. Atrophy is comparatively easy to diagnose in this condition since comparison with adjacent muscles is available. The upper and middle thirds of the muscle belly are affected to a greater extent helping distinguish this condition from other diseases that affect the entire muscle belly, such as myopathies and denervation injuries. The muscle bulk is usually normal in other conditions causing restriction of hip movements such as inflammatory pathologies like iliopsoas tendinitis and abscesses and the muscle maybe enlarged in the case of a muscle contusion.

The intramuscular fibrous cord appears as a linear band of low signal intensity on both T1 and T2 weighted images and is best appreciated with fat suppressed images. The cord maybe thick or thin and is responsible for retraction of the muscle giving rise to secondary features. Secondary MRI findings include medial retraction of the distal body and tendon of the gluteus maximus with depression at the myotendinous junction (which may produce a dimple on surface examination), posteromedial retraction of the iliotibial tract and abnormal external rotation of the affected hip.

Imaging plays an important role in characterizing the extent and severity of the disease. It is important to exclude the involvement of the gluteus medius and minimus before release of a gluteus maximus contracture as this could lead to near complete loss of abductor function on the affected side and a Trendelenburg gait could develop after surgery.

Treatment

Conservative therapy for this syndrome includes physiotherapy, massage, shortwave diathermy and application of hot packs over the buttocks [13]. However, stretching exercises are not very useful in long standing cases with already developed contractures (seen as a fibrous cord on imaging) [12]. Surgical release of the fibrous cord (contracture) is thus, the treatment of choice in most cases. This maybe coupled with fasciotomy of the iliotibial tract or Z-plasty lengthening of the affected muscle [1,7]. The complications of surgery include hematoma formation, wound infection, keloid formation, etc. Excessive release or injury to the gluteus medius may also produce a Trendelenburg's gait post-operatively [13]. Thus, exclusion of associated gluteus medius contracture before surgery and careful surgical technique is very important.

Conclusion

In conclusion, gluteus maximus contracture syndrome is an uncommon but debilitating condition which needs to be diagnosed and corrected early. Imaging studies are useful in evaluating the extent of disease and muscles involved as well as in planning of surgery.

TEACHING POINT

Gluteus maximus contracture, albeit uncommon, is a debilitating disease that can lead to severe morbidity and although, clinical diagnosis is not very difficult, imaging plays an important role in aiding diagnosis and characterizing the extent of disease involvement. Detection of muscle atrophy and demonstration of an intramuscular fibrous band on imaging strengthens the diagnosis and early treatment can lead to significant functional improvement and prevent irreversible changes.

REFERENCES

1. Hang YS. Contracture of the hip secondary to fibrosis of the gluteus maximus muscle. *J Bone Joint Surg Am.* 1979 Jan; 61(1):52-5. PMID: 759436.
2. Chung Dc, Ko YC, Pai HH. A study on the prevalence and risk factors of muscular fibrotic contracture in Jia-Dong Township, Pingtung County, Taiwan. *Gaoxiong Yi Xue Ke Xue Za Zhi.* 1989 Feb;5(2):91-5. PMID: 2786573.
3. Liu Y, Wang Y, Xue J, Lui PP, Chan K. Arthroscopic gluteal muscle contracture release with radiofrequency energy. *Clin Orthop Relat Res.* 2009 Mar;467(3):799-804. PMID: 18975040.
4. Sun X. An investigation on injectional gluteal muscle contracture in childhood in Mianyang City. *Zhonghua Liu Xing Bing Xue Za Zhi.* 1990 Oct; 11(5):291-4. PMID: 2261618.
5. Peiro A, Fernandez CI, Gomar F. Gluteal fibrosis. *J Bone Joint Surg Am.* 1975 Oct;57(7):987-90. PMID: 1184648.
6. Shanmugasundaram TK. Post-injection fibrosis of skeletal muscle: a clinical problem. A personal series of 169 cases. *Int Orthop.* 1980;4(1):31-7. PMID: 7399778.
7. Ko YC, Chung DC, Pai HH. Intramuscular-injection-associated gluteal fibrotic contracture and hepatitis B virus infection among school children. *Gaoxiong Yi Xue Ke Xue Za Zhi.* 1991 Jul;7(7):358-62. PMID: 1875457.
8. Jackson AM, Hutton PA. Injection-induced contractures of the quadriceps in childhood: A comparison of proximal release and distal quadricepsplasty. *J Bone Joint Surg Br.* 1985 Jan; 67:97-102. PMID: 3968154.

9. Chen CK, Yeh L, Chen CT, Pan HB, Yang CF, Resnick D. Contracture of the deltoid muscle: imaging findings in 17 patients. *AJR Am J Roentgenol.* 1998 Feb;170(2):449-53. PMID: 9456962.
10. Fernandez de Valderrama JA, Esteve de Miguel R. Fibrosis of the gluteus maximus: a cause of limited flexion and adduction of the hip in children. *Clin Orthop Relat Res.* 1981 May;(156):67-78. PMID: 7226666.
11. Gao GX. Idiopathic contracture of the gluteus maximus muscle in children. *Arch Orthop Trauma Surg.* 1988;107(5):277-9. PMID: 3178441.
12. Sirinelli D, Oudjhane K, Khouri N. Case report 605: Gluteal amyotrophy: a late sequela of intramuscular injection. *Skeletal Radiol.* 1990;19(3):221-3. PMID: 23333564.
13. Zhao CG, He XJ, Lu B, Li HP, Wang D, Zhu ZZ. Classification of gluteal muscle contracture in children and outcome of different treatments. *BMC Musculoskelet Disord.* 2009 Apr 7;10:34. PMID: 19351391.
14. Lieberman DE, Raichlen DA, Pontzer H, Bramble DM, Cutright-Smith E. The human gluteus maximus and its role in running. *J Exp Biol.* 2006 Jun;209(Pt 11):2143-55. PMID: 16709916.
15. Zhao CG, He XJ, Lu B, Li HP, Kang AJ. Increased expression of collagens, transforming growth factor-beta1, and -beta3 in gluteal muscle contracture. *BMC Musculoskelet Disord.* 2010 Jan 25;11:15. PMID: 20100316.
16. Chen CK, Yeh L, Chang W, Pan HB, Yang CF. MRI diagnosis of contracture of the gluteus maximus muscle. *AJR Am J Roentgenol.* 2006 Aug;187(2):W169-74. PMID: 16861506.
17. Ye B, Zhou P, Xia Y, Chen Y, Yu J, Xu S. New minimally invasive option for the treatment of gluteal muscle contracture. *Orthopedics.* 2012 Dec;35(12):e1692-8. PMID: 23218623.
18. Ni B, Li M. The effect of children's gluteal muscle contracture on skeleton development. *Sichuan Da Xue Xue Bao Yi Xue Ban.* 2007 Jul;38(4):657-9, 677. PMID: 17718434.
19. Cai J, Gan L, Zhen H, Li H. Iliac hyperdense line: a new radiographic sign of gluteal muscle contracture. *Pediatr Radiol.* 2005 Oct;35(10):995-7. PMID: 15965678.
20. Li Q, Lingyan Z, Yan L, Yulan P. The role of ultrasonography in the diagnosis of gluteal muscle contracture. *Skeletal Radiol.* 2011 Feb;40(2):215-21. PMID: 20140430.

FIGURES

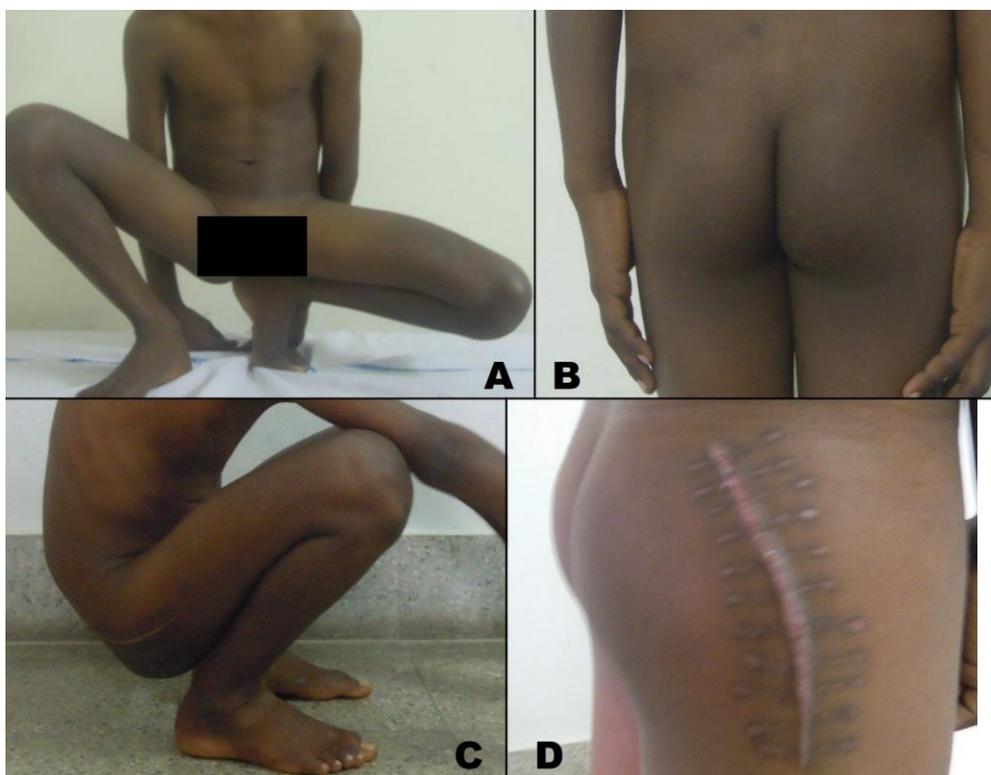


Figure 1: 10 year old male with bilateral gluteus muscle contracture syndrome. A) Pre-operative clinical photograph showing an abducted-externally rotated bilateral hip deformity while squatting B) Pre-operative clinical photograph showing flattened buttocks C) 6 weeks post-operative clinical photograph showing improvement of deformity in the squatting position D) 6 weeks post-operative clinical photograph showing the healed surgical scar.

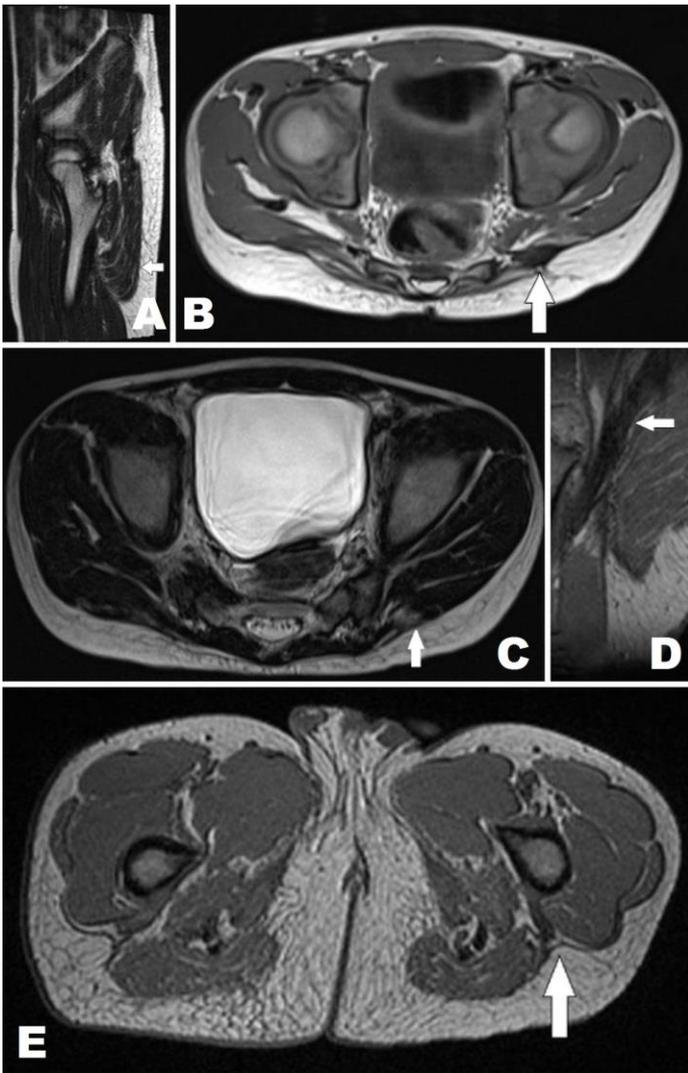


Figure 2 (left): MRI (1.5T, standard body matrix) images of the pelvis of a 10 year old male with bilateral gluteus muscle contracture syndrome. A) T2 (TR/TE 1500/127, 1 mm) sagittal image showing atrophy of the gluteus maximus (arrow) B) T1 (TR/TE 772/12, 3 mm) axial image showing the low intensity fibrous band (arrow) within the left gluteus maximus C) T2 (TR/TE 1500/127, 1 mm) axial image showing atrophy and a fibrous band (arrow) within the left gluteus maximus D) T1 curved MPR image showing the fibrous band (arrow) in its entire extent E) T1 (TR/TE 1800/12, 1 mm) axial image showing medial retraction of the distal body and tendon of the gluteus maximus (arrow) and posteromedial retraction of the iliotibial tract.

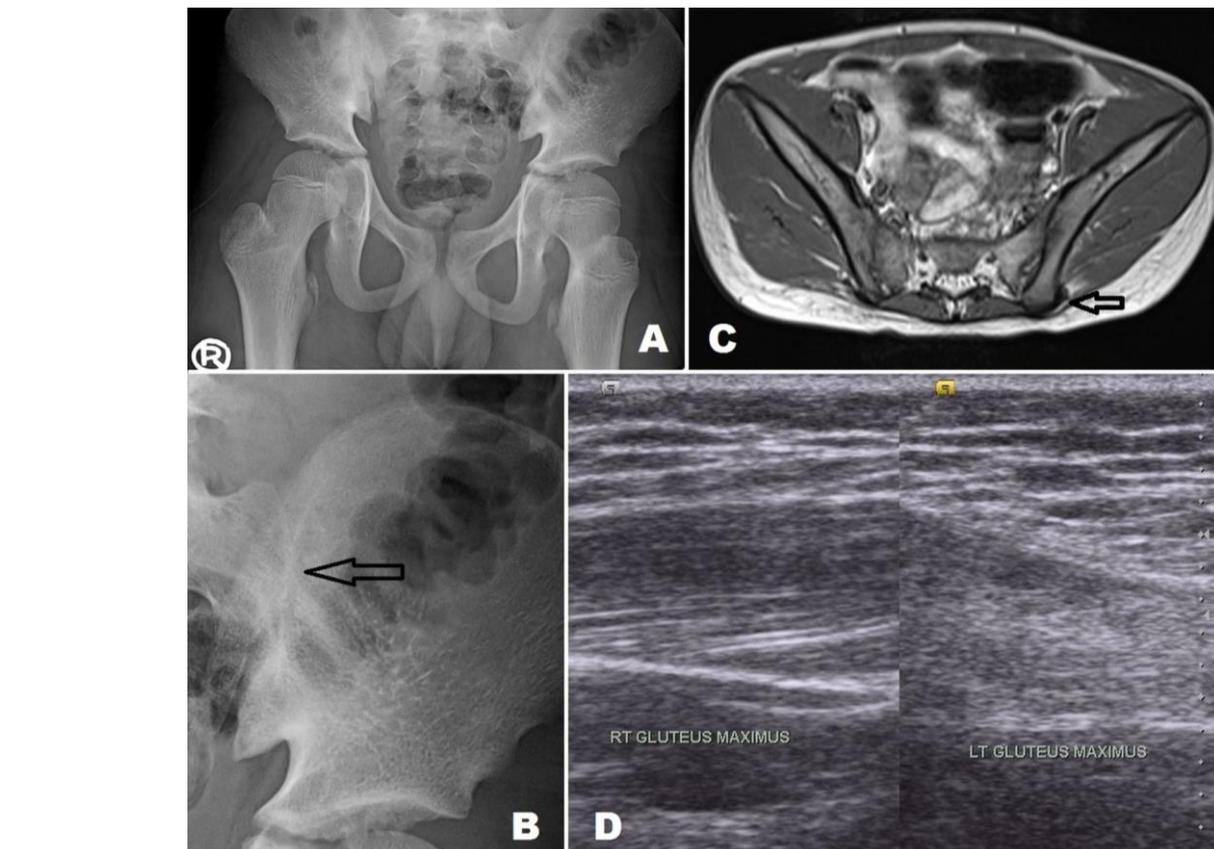


Figure 3 (bottom): 10 year old male with bilateral gluteus muscle contracture syndrome. A) Plain AP radiograph (Siemens 600 mA, 80 kV, 20 mAs) of pelvis showing a subtle vertical curved line along the lateral iliac cortex adjacent to the left sacro-iliac joint (arrow). B) Magnified image of the AP radiograph showing the line better (arrow). C) T1 weighted axial MR (1.5T, standard body matrix) image showing the deformity of the lateral iliac cortex at the site of attachment of gluteus maximus (arrow) which is the cause of the vertical line on plain radiographs. D) USG (Siemens Acuson X300, 8 MHz, linear transducer) comparison of both glutei maximus showing increased echoreflexivity of the left one.

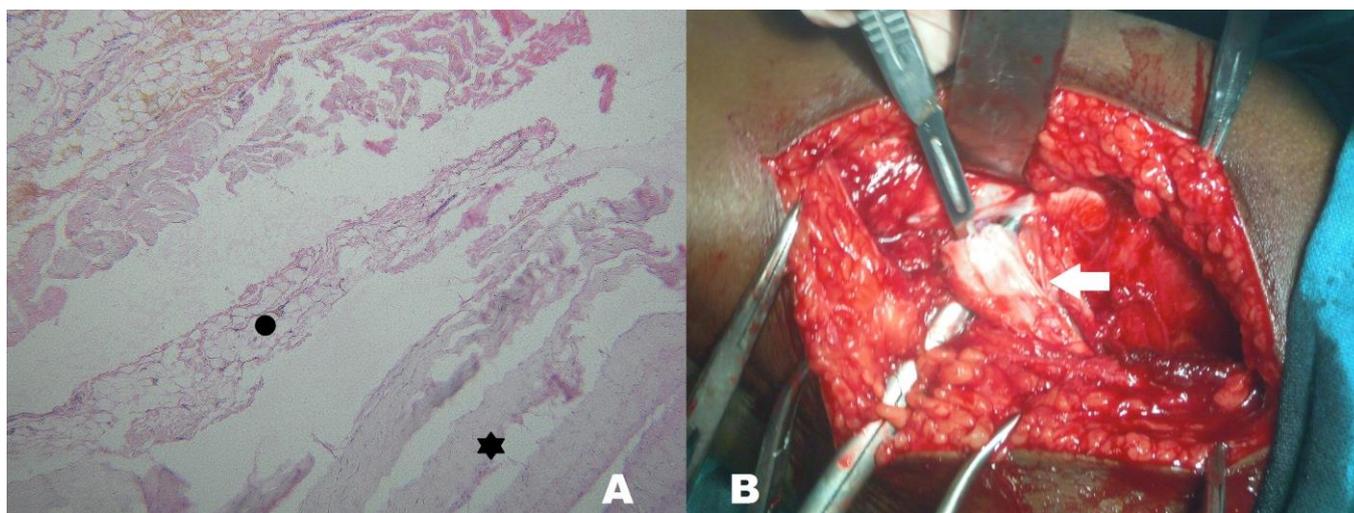


Figure 4: 10 year old male with bilateral gluteus muscle contracture syndrome. A) Photomicrograph of a slide with Hematoxylin-Eosin stained histopathologic specimen showing presence of fibrous tissue (*) and adipose tissue (.) within the muscle B) Intra-operative photograph showing the fibrous band.

Incidence	1 – 2.5%
Etiology	Multiple intramuscular injections; congenital
Demographics	School going children; 6-18 years, boys more than girls
Clinical examination	Squatting test – abduction-external rotation deformity. Active flexion test – limited range of hip flexion.
Plain Radiograph	Vertical pelvic hyperdense line along lateral iliac cortex
Ultrasonography	Increased echogenicity of muscle with intramuscular fibrous band
MRI	Muscle atrophy, intramuscular fibrous band, medial retraction of the distal belly and tendon, posteromedial retraction of the iliotibial tract at attachment
Treatment	Surgical release of contracture with or without Z-plasty and muscle lengthening; physiotherapy.
Prognosis	Good if diagnosed and corrected early.

Table 1: Summary table for Gluteus Maximus Contracture Syndrome

	Plain Radiography	Sonography	MRI
Gluteus Maximus Contracture	Iliac hyperdense line	Increased echoreflectivity and muscle thinning	Muscle atrophy, intramuscular fibrous band, medial retraction of tendon; upper and middle third more affected.
Restricted hip abduction due to inflammatory conditions (iliopsoas tendinitis, abscess)		Normal gluteus maximus, primary cause maybe diagnosed	Normal gluteus maximus, primary cause maybe diagnosed
Denervation Injury		Thinning of muscle belly	Muscle atrophy, entire muscle involved
Muscle injury (contusion)	May have associated fractures	Focal hyperechoic within muscle belly	T2 hyperintense focus, T1 hyperintense hematoma.

Table 2: Differential diagnosis table for Gluteus Maximus Contracture Syndrome

ABBREVIATIONS

MR - Magnetic Resonance
MRI - Magnetic Resonance Imaging
USG – Ultrasonography

KEYWORDS

Gluteus maximus; contracture; MRI pelvis; hip abduction deformity; congenital

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