Congenital Heart Disease in Adolescents With Gluteal Muscle Contracture

Tian You, MD, Xin-tao Zhang, MD, Zhen-gang Zha, MD, and Wen-tao Zhang, MD

INTRODUCTION

Gluteal muscle contracture (GMC), also named “injection contracture,” is a clinical syndrome characterized by gait abnormality and limb dysfunction including restriction of adduction and internal rotation of the hip joint. It was first found by Valderrama at a scientific meeting of the British Orthopedic Association in London in 1969. Its pathologic change typically presents with fibrosis and contracture of the gluteus and its fascia. The knees cannot be brought together and are separated in a frog-leg position. As younger patients with GMC continue to develop and grow, pathologic changes increase in prevalence and severity including leg length discrepancy, pelvic oblique, compensatory scoliosis, and, in severe cases, bilateral dislocation of the hip joints. Now, arthroscopic release, a minimally invasive surgery which was reported by Zhang et al for the first time, has become the gold standard of treatment in GMC patients.

It is reported that GMC is related to benzyl alcohol. Compared with developed countries, GMC is more widely reported in China. So far, numerous studies have suggested that GMC usually was associated with repeated intramuscular injection into the gluteal region during childhood. However, it remains unclear that the reasons why these children are weak and have no choice to accept repeated intramuscular injection.

The paper reveals that CHD children are prone to catching cold, repeated respiratory tract infection, or pneumonia and have to take frequently intramuscular injection which use benzyl alcohol as a solvent for penicillin in some regions of China in the 1970s and 1980s, leading to degeneration, necrosis and fibrosis of the gluteal muscles and fascia, as well as serious limitation of hip movements.

PATIENTS AND METHODS

We identified 4 GMC adolescents with CHD from January 2013 to March 2014. The duration of symptoms ranged from 6 to 10 years. All had been unable to improve with functional exercise. There were 2 males and 2 females with age ranging from 14 to 17 years (Table 1). All patients provided informed consent for the operation and followed our rehabilitation program. This study was approved by the Institutional Review Board of our institution. All patients had a history of injections around the gluteal region. Although 3 of 4 patients (patients 1, 3, and 4) had limitations with activities of daily living, they believed their activity levels were lower than those of healthy people. Three of 4 patients (patients 1, 3, and 4) had an abnormal gait with out-toe walking, and could not crouch with both knees close to each other or sit down with their legs crossed. All patients had a snapping sound during rotation of the hip. Patient 1 was diagnosed as CHD for the first time, while the other 3 had been cured yet. Patient 4 had received open surgery for GMC but dissatisfied result. Two representative patients (Patients 1 and 4) were selected for further discussion of their clinical and radiographic findings (Table 2).

Each author certifies the study had approved, by Ethics committee of Peking University Shenzhen Hospital, for the
reporting of these cases and that all investigations were conducted in conformity with ethical principles of research.

**CASE REPORTS**

**Case 1**

Patient 1 was a 16-year-old girl who had bilateral diseases with gluteal muscle atrophies, abnormal gait with out-toe walking, dimpling of skin around the gluteal regions, snapping hips, adduction and internal rotation dysfunction of hips, Ober’s sign. The most thing affected her was adduction and internal rotation dysfunction of hips, which made her not to do competitive sports (actually, it was mainly influenced by CHD which was diagnosed later.) like other healthy girls. 3D reconstruction CT showed that her gluteus maximus and gluteus medius were obviously dysplastic and part of them turned to contractile bands. We heard a systolic ejection murmur at the second left intercostal space in preoperative examinations. Furthermore, Chest X-ray revealed pulmonary vascularity, cardiac enlargement and aortic knob shrinking. Finally, cardiac ultrasound proved the diagnosis of ASD. Because of this, we sent her to cardiosurgery department for atrial septal defect closure and partial anomalous pulmonary venous drainage correction on beating heart. The arthroscopic release and rehabilitation program for GMC were operated 110 days later, when she was completely rehabilitated from cardiosurgery. Three months later, the patient gained a satisfactory result without complications, snappings, and dysfunctions of hip (Figure 1).

**TABLE 1. Clinical Characteristics of the Patients**

| Patient | Age (years) and Sex | Symptom and Sign | CHD   | Open Surgery of GMC |
|---------|---------------------|------------------|-------|---------------------|
| 1. Yue  | 16, female          | Gluteal muscle atrophy, abnormal gait with out-toe walking, dimpling of skin around the gluteal region, snapping hip, adduction and internal rotation dysfunction of hip, Ober’s sign | ASD   | No                  |
| 2. Huang| 14, female          | Snapping hip     | VSD, cured | No               |
| 3. Xu   | 14, male            | Gluteal muscle atrophy, abnormal gait with out-toe walking, dimpling of skin around the gluteal region, snapping hip, adduction and internal rotation dysfunction of hip, Ober’s sign | TOF, cured | No               |
| 4. Yang | 17, male            | Gluteal muscle atrophy, abnormal gait with out-toe walking, dimpling of skin around the gluteal region, snapping hip, adduction and internal rotation dysfunction of hip, Ober’s sign | PDA, cured | Yes, 4 years ago |

ASD = atrial septal defect, CHD = congenital heart disease, GMC = gluteal muscle contracture, PDA = patent ductus arteriosus, TOF = tetralogy of fallot, VSD = ventricular septal defect.

**TABLE 2. Radiographic Findings of the Patients**

| Patient | Age (Years) and Sex | Pelvis CT | Chest X-Ray | Cardiac Ultrasound |
|---------|---------------------|-----------|-------------|-------------------|
| 1. Yue  | 16, female          | Part of gluteus maximus and gluteus medius atrophy and turning to contractile bands | Pulmonary ascularity, cardiac enlargement, aortic knob shrinking | Ventricular septal defect, right atrium and ventricle enlargement, pulmonary artery dilation, left ventricular ejection (LVEF), and fraction shortening (FS) are normal |
| 2. Huang| 14, female          | Part of gluteus maximus and gluteus medius atrophy and turning to contractile bands | Normal, mild scoliosis | Normal |
| 3. Xu   | 14, male            | Part of gluteus maximus and gluteus medius atrophy and turning to contractile bands | Normal, there are four metal sutures in sternal area | Pulmonary artery blood flow speeding up, left ventricular ejection (LVEF), and fraction shortening (FS) are normal unavailable |
| 4. Yang | 17, male            | Part of gluteus maximus and gluteus medius atrophy and turning to contractile bands | Normal | unavailable |

3D-CT = three-dimensional computed tomography, CT = computed tomography, FS = fraction shortening, LVEF = left ventricular ejection fraction.
FIGURE 1. (A) A 3D reconstruction CT shows the gluteal muscles of Patient 1, a 16-year-old woman who had GMC with severe gluteus maximus and gluteus medius atrophy. (B) and (C) show the contracture bands (white arrow) in left and right side, respectively. (D) Cardiac ultrasound demonstrates ASD. (E) Chest X-ray reveals pulmonary ascularity, cardiac enlargement and aortic knob shrinking.

FIGURE 2. (A) A 3D reconstruction CT shows the gluteal muscles of Patient 4, a 17-year-old man who had GMC with mild gluteus maximus and gluteus medius atrophy. (B) and (C) show the contracture bands (white arrow) in left and right side, respectively. (D) Chest X-ray reveals normal.
Case 2
Patient 4 was a 17-year-old boy who had bilateral diseases with gluteal muscle atrophies, abnormal gait with out-toe walking, dimpling of skin around the gluteal regions, snapping hips, adduction and internal rotation dysfunction of hips, Ober’s signs. The most things affected him was adduction and internal rotation dysfunction of hips, as well as snapping hips. Due to a failed open surgery before, he decided to choose the minimally invasive surgery. When he was very young, he was taken ligation of patent ductus arteriosis (the detailed data of that surgery were unavailable). Physical examinations and chest X-ray were normal. 3D reconstruction CT showed that his gluteus maximus and gluteus medius were obviously dysplastic and part of them turned to contractile bands. Then, we implemented the arthroscopic release and rehabilitation program of GMC. Three months later, the patient gained a satisfactory result without complications, snappings, and dysfunctions of hip (Figure 2).

DISCUSSION
We present a series of 4 GMC patients with CHD, including ASD, VSD, TOF, and PDA respectively. Accordingly, we suggest that some GMCs are related to CHDs, or rather are derived from CHDs. To be specific, children suffered from CHDs are easy to acquire respiratory infections (nearly one-quarter to one-third\textsuperscript{13}), which may be result in frequent use of benzyl alcohol as a dissolvent for penicillin for intramuscular injections in some regions of China in 1970s–1980s, as several researchers have suggested that benzyl alcohol is the main cause of GMC.\textsuperscript{11–12}

This series is limited by its small size relating to the decreasing of GMC for abandoning benzyl alcohol as a dissolvent. Patient 1 was admitted to hospital for releasing GMC, however, ASD was found in preoperative examinations. This warns that surgeons should attach more importance to the cardiac examination, especially identifying the real cause of “cannot do competitive sports.” When CHD is repaired and physical condition is fully recovered, it is time to release GMC. At last, the patient acquired an excellent outcome without complications. Patient 4 was hospitalized in order to cure GMC through a noneffective open surgery. When he was very young, he was taken for releasing GMC, and part of them turned to contractile bands. Then, we implemented the arthroscopic release and rehabilitation program of GMC. Three months later, the patient gained a satisfactory result without complications, snappings, and dysfunctions of hip (Figure 2).

CONCLUSIONS
GMC with CHD is uncommon. Although there are no exact cause–result relationship between the 2 diseases, we propose 3 recommendations: (1) as to CHD children, in addition to treating the primary disease as early as possible, it is essential to decreasing the use of intramuscular injection, and (2) paying more attention to cardiac examination especially cardiac ultrasound in perioperative period, and (3) taking 3D-CT to reconstruct gluteal muscles for observing contracture bands clearly in preoperation.

REFERENCES
1. Liu GH, Cao FQ, Yang SH, Zhu JF. Factors influencing the treatment of severe gluteal muscle contracture in children. J Pediatr Orthop B. 2011;20:67–69.
2. Fernandez de Valderrama JA. A cause of limited flexion and adduction of the hip in children. J Bone Joint Surg (Br). 1970;52:179.
3. Fu D, Yang S, Xiao B, et al. Comparison of endoscopic surgery and open surgery for gluteal muscle contracture. J Pediatr Orthop. 2011;31:e38–e43.
4. Liu YJ, Wang Y, Xue J, et al. Arthroscopic gluteal muscle contracture release with radiofrequency energy. Clin Orthop Relat Res. 2009;467:799–804.
5. Dini GM, Ferreira LM. Capsular contracture in gluteal implant patients. Plast Reconstr Surg. 2006;117:1070–1071.
6. Zhang WT, Wang Y, Wang ZG, et al. Treatment of the gluteal muscle contracture after intramuscular injections and snapping hip with arthroscopic release. Chin J Clin Rehabil. 2002;6:1758–1759.
7. Cui JC, Wang WC, Wu B, Wang SY. Release of gluteal muscle contracture by radiofrequency under arthroscopy. Zhong Nan Da Xue Xue Bao Yi Xue Ban. 2008;33:274–276.
8. 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. Gao xiong Yi Xue Ke Xue Za Zhi. 1989;5:91–95.
9. 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;156:67–68.
10. Huang Y, Li J, Lei W. Gluteal muscle contracture: etiology, classification and treatment. Chin J Orthop. 1999;19:106–108.
11. Peng M, Zhou Z, Zhou X. Epidemiology of gluteal muscle contracture in Si Chuan Province. Chin J Pediatr Surg. 1989;10:356–358.
12. Sirinelli D, Oudjhane K, Khouri N. Case report 605: gluteal atrophy: a late sequel of intramuscular injection. Skeletal Radiol. 1990;19:221–223.
13. Rudolph AM. Diagnosis and treatment: respiratory distress and cardiac disease in infancy. Pediatrics. 1965;35:999–1002.