บทคัดย่อ
ไลโปมัยอีโลซีล (lipomyelocele) พบได้น้อยมากในทารกแรกเกิด พบได้ตั้งแต่ 0.3 – 0.6 ต่อทารกแรกเกิด 10,000 คน แพทย์ผู้ให้การรักษารายงานผู้ป่วยเด็กเพศหญิงอายุ 6 ปี ได้รับการรักษาที่มีไลโปมัยอีโลซีล โดยไม่มีอาการแสดงใดๆ ตั้งแต่แรกเกิดมาเป็นเวลา 6 ปี หลังจากนั้นเริ่มมีอาการผิดปกติทางระบบประสาท จากการทบทวนประวัติผู้ป่วยรายนี้ พบว่า มีรอยบุ๋มที่มีหนังบริเวณกลางหลังตั้งแต่แรกเกิดและไม่ได้รับการตรวจรักษา ตอนอายุ 7 ปี ผู้ป่วยได้รับการรักษาด้วยการผ่าตัดเอาก้อนไขมันออกและซ่อมแซมเยื่อบุรู แต่อย่างไรก็ตามการรักษาไม่สามารถแก้ไขอาการผิดปกติทางระบบประสาทให้กลับคืนได้

คําสั่งห์: สไปนอลดิสราฟิซิ่ม ไลโปมัยอีโลซีล ซิโนดิมเพิล

บทวิจารณ์

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Skin dimple, more than a pit

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Abstract

Lipomyelocele is an uncommon condition in neonates with a low prevalence of 0.3 – 0.6 per 10,000 live births. We report a case of 6-year-old girl with lipomyelocele who had no neurological deficit for 6 years. Afterward, she presented with progressive neurological deficits. Her history was revealed that she had a typical cutaneous marker which was abandoned since birth. Finally, she received a surgical removal of a lipomatous mass and a dural reparation at the age of 7 years old. Unfortunately, her neurological deficits cannot be recovered.

Keywords: spinal dysraphism, lipomyelocele, skin dimple

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Introduction

Neural tube defects are congenital anomalies of neurulation in embryogenesis with a wide range of clinical manifestations. Lipomyelocele is one of the neural tube defects which are classified in closed spinal dysraphism (occult spinal dysraphism). It usually presents with cutaneous markers or neurological deficits secondary to tethered spinal cord. Mainstay of treatment is surgery, which aim to prevent further neurological deficits. We report a case of delayed diagnosed lipomyelocele with review literatures in aspect of treatment and outcome.

Case report

A 6-year-old girl presented with a progressive foot drop on her right leg, numbness of her right foot and episodic urinary urgency for 6 months. Physical examinations revealed muscle strength of 3/5 of right tibialis anterior muscle, extensor hallucis longus muscle, and gastrocnemius muscle. She also had a hyporeflexia of the right ankle and decreased pin-prick sensation of the lateral aspect of the right foot. She had a skin dimple which located in the midline of lumbrosacral region, her mother claimed that it presented at birth.

Magnetic Resonance Imaging (MRI) of the lumbosacral spine showed spina bifida occulta from L4 through S5 level. The corresponding spinal canal was capacious and posterior scalloping of L4 and L5 vertebrae were detected. A 2.2×1.6×6.2 cm subcutaneous fatty mass was seen at the covering skin that represents lipoma. It extended toward from the subcutaneous plane through the defects into the spinal canal. It inserted into the dorsal surface of the tethered spinal cord which terminated at upper S1 level (Figure 1).

She was diagnosed with lipomyelocele with no associated abnormalities. Removal of the lipomatosus mass, untethering the spinal cord, and duroplasty with autologous fascial graft were done. After the operation, she had more weakness in the former described muscles, muscle strength of the affected limb become 1/5. She developed neurogenic bladder resulting from retained catheter since the operation.

Discussion

Neural tube defects can affect cranium, spine or both as in the following: 1. Cranial defects including anencephaly, exencephaly, and encephalocele, 2. Open spinal dysraphism (spina bifida aperta) is characterized by herniation of the nervous tissue through the defect in the spinal column including myelomeningocele and meningocele, 3. Closed spinal dysraphism (spina bifida occulta) is characterized by unexposed nervous tissue. Lipomyelocele is a common subtype of the closed spinal dysraphism. The prevalence of all types of spinal dysraphism occur at a frequency of 0.5 to 8 per 1,000 live births while the prevalence of lipomyelocele has been range from 0.3 to 0.6 per 10,000 live births.
Risk factors of spinal dysraphism are ethnic groups, nutritional deficiency, and maternal diabetes. First, prevalence of spinal dysraphism in Hispanic group is 3.26 per 10,000 live births while prevalence of spinal dysraphism in non-Hispanic whites and non-Hispanic blacks are 2.57 and 2.07 per 10,000 live births, respectively. Second, folic acid deficiency has been reported as a strong association with the open spinal dysraphism. There are also several reports of a significant higher prevalence of open spinal dysraphism in neonates which born to pregnant women who take certain antiepileptic drugs which affect in folic acid metabolism. Third, maternal diabetes has been associated with increased incidence of the syndrome of caudal regression without exactly known mechanism. Conversely, there are no significant risk factors which closely correlated with spinal dysraphism.

Lipomyelocele results from abnormal embryogenesis during primary neurulation. According to premature dysjunction of neuronal ectoderm and epidermal ectoderm, these abnormal development allow mesenchyme to attach to the developing neural tube. This mesenchyme is induced by dorsal surface of neural tube to form fatty tissue, and prevents proper neurulation. Closed spinal dysraphism are commonly accompanied by cutaneous markers such as subcutaneous masses, tails, capillary hemangioma, dimples, and hairy nevus in 43-95% of cases. In Natarajan cases series reported that merely 40% of patients with lipomas without a dural defect had cutaneous markers while all of patients with lipomas with dural defect had cutaneous markers. These cutaneous markers can be used as indicators for further investigation in asymptomatic neonates.

Progressive neurological deficits, urinary deficits, orthopedic deformities and the sequelae of tethered cord has been found to increase with age because of progressive clonus tethering and injury to nervous tissue. Hoffman et al. observed that 62.5% of patients were neurologically asymptomatic before 6 months of age while only 29.3% of patients who presented after 6 months of age were asymptomatic. Among the symptomatic patients, they can present as weakness or hypalgesia of one or both lower extremities, gait abnormalities, sphincter disturbance, scoliosis, limb length discrepancy, foot deformities, or back and leg pain.

Treatment strategies of lipomyelocele mainly are surgical interventions and rehabilitation in deficit cases. Surgery in these groups of patients consisted of removal of lipomatous mass, avoid injuring any nerve roots or clonus medullaris that might course close to dural defect, and primary dural closure or duroplasty in large defects. Complications can be divided into two types: wound complications and neurological complications. Incidence of wound complications occurred in 10-30% of patients included wound infection, flap necrosis, and CSF leakage. Neurological deterioration after surgery can be transient or permanent neurological deficits. Temporary weakness or urinary deficits reported in 3.4-7.5% after the operation. Permanent neurological deficits has been found 3-10% which could be influenced by surgical treatment, incomplete release of tethering, or retethering. Retethering occurred 3.45-20% of the patients after follow-up for 3-8 years. Treatment outcomes in one case reported that 44.83% of patients improved, 18.97% stabilized deficits, 3.45% deteriorated, and 32.76% of patients were asymptomatic, same as presentation. Preservation or improvement of neurological outcomes is better when patients presented and underwent surgery before 2 years of age.

In conclusion, we report a case of lipomyelocele which was diagnosed by presentation of neurological deficits. Despite this patient had a typical cutaneous marker at birth, which we; as a physician, should aware and promptly investigate for spinal dysraphism. We had missed a 6-year-period of a curative treatment for her. Our purpose is to emphasize the important of a skin dimple in neonate is not just a pit.
บทคัดย่อ

"โฟกัส T25" เป็นโปรแกรมการออกกำลังกายระดับหนัก ผลิตโดยบริษัท บีชบอดี้ โดย ชอน ที่ ซึ่งเป็นผู้สร้างโปรแกรม อินแซนิตี้ ที่เป็นที่นิยมอย่างมากเช่นกัน แต่ใช้เวลานานกว่าเล็กน้อย "โฟกัส T25" เป็นโปรแกรมที่ใช้เวลา 25 นาที ที่มีการฝึกกายภาพประมาณ 5 นาที ภายหลังออกกำลังกาย สิ้นสุด ที่ ชอน ที่ ใช้คำว่า "โฟกัส" มาหน้า เพราะเป็นโปรแกรมการออกกำลังกายที่เน้นท่าและจังหวะของการออกกำลังกาย ที่ทำให้สร้างกล้ามเนื้อและลดไขมันได้มากที่สุด ในเวลา 25 นาที โดยสรุปแล้ว "โฟกัส T25" มีผลเพิ่มความแข็งแรง พลังและความอดทนของกล้ามเนื้อทั้งร่างกาย เพิ่มความสดของระบบหัวใจและระบบหายใจ เพิ่มความแข็งแรงสมดุลการทรงตัว ความยืดหยุ่น และการควบคุมอุณหภูมิร่างกาย ดังนั้น "โฟกัส T25" จึงน่าจะมีบทบาทในการสร้างเสริมสุขภาพได้อย่างไรก็ตามหากทำไม่ถูกวิธีอาจทำให้เกิดอันตรายได้ เพราะมีระดับการออกกำลังกายที่หนักมาก และมีการกระโดดต่อเนื่องกันเป็นเวลานาน หากร่างกายไม่พร้อม อย่างทุกส่วนของร่างกาย เช่น กล้ามเนื้อและเอ็น อาจทำให้เกิดอันตรายได้ บทความนี้จึงได้สรุปประโยชน์ของ "โฟกัส T25" และข้อควรระวัง รวมทั้งการหยุดการออกกำลังกาย เพื่อให้ผู้ออกกำลังกายได้ประโยชน์จากการออกกำลังกายแบบโปรแกรม "โฟกัส T25" ได้อย่างเต็มที่โดยไม่เกิดอันตรายแก่ร่างกาย

คำาสำคัญ: การออกกำลังกายระดับหนัก ความแข็งแรง ความอดทน การทรงตัว การควบคุมอุณหภูมิ

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Physiological responses to “Focus T25”

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Abstract

“Focus T25” is a high-intensity exercise program. It is produced by beach body and led by world renowned personal trainer Shaun-T. Shaun-T was the program creator of the incredibly popular Insanity program which is a bit longer in duration. “Focus T25” takes around 25 minutes with 2-5 minutes of stretching after 25-minute workout. The word “focus” was used because Shaun T has chosen the most effective exercise combinations that will build and burn a large number of muscles and fat in the quickest time possible within 25 minutes. Overall, “Focus T25” increases muscular strength, power and endurance, cardiorespiratory endurance, speed, balance and flexibility. In addition, it improves body temperature regulation. Therefore, it could be benefit for health promotion. However, if we do it wrong we may get injury from its high intensity and jumping. If the exercisers are not fit enough we may experience muscle and tendon injuries, fatigue, joint pain, faint or heart failure. Beneficial physiological effects of “Focus T25” following by precaution and termination of the exercise are summarized. This may help exercisers gain full benefit from “Focus T25” with safety.

Keywords: high intensity exercise, strength, endurance, balance, weight reduction

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Introduction

At this moment in Thailand, there is no exercise program popular than “FocusT25” for anyone who loves the intense weight and cardio workouts. “Focus T25” is produced by beach body and led by world renowned personal trainer Shaun-T. Shaun-T was the program creator of the incredibly popular Insanity program which is a bit longer in duration. “Focus T25” takes around 25 minutes with 2-5 minutes of stretching after 25-minute workout. The word “focus” was used because Shaun T has chosen the most effective exercise combinations that will build and burn a large number of muscles and fat in the quickest time possible within 25 minutes. The exercisers do not stop once the DVD starts up.

However, “Focus T25” has two sides; benefit if we do it right but dangerous if we do it wrong. So I start from summarizing the beneficial physiological effects of “Focus T25” following by precaution and termination of the exercise. This may help exercisers gain full benefit from “Focus T25” with safety.

Beneficial physiological effects

World Health Organization recommended that vigorous-intensity activities at least 3 days per week; and activities that strengthen muscle and bone at least 3 days per week provides greater health benefit. “Focus T25” is a high-intensity exercise program. It involves high impact moves that engage all areas of exerciser’s core, major muscle groups like quadriceps, calves and hamstrings, trunk and the upper body. Overall, “Focus T25” increases muscular strength, power and endurance, cardiorespiratory endurance, speed, balance and flexibility. In addition, it improves body temperature regulation. Therefore, it could be benefit for health promotion.

In order to have these capacities, our bodies need energy supply. Only one molecule that can immediately provide energy is adenosine triphosphate (ATP). However, the body cannot easily store ATP (and what is stored gets used up within a few seconds), it is necessary to continually synthesize ATP during exercise.

Generally, the two major ways the body converts nutrients to energy are (Figure 1): aerobic metabolism (with oxygen) and anaerobic metabolism (without oxygen). Carbohydrate (CHO) is the main nutrient that provides energy during a high-intensity exercise via anaerobic glycolysis or aerobic pathway, while fat can provide energy during a low-intensity exercise for long periods of time via aerobic pathway (Figure 2). Proteins are generally used to maintain and repair body tissues, and are not normally used to power muscle activity. However, Thai healthy sedentary subjects relied more on CHO than fat during all (Figure 3). The greater daily proportional CHO intake in Thai subjects than that in white subjects may be responsible for the greater CHO utilization. Thus, not only the intensity and duration of the exercise determine which method gets used but also the diet.

In addition, “Focus T25” is the program that follows the overload training principle. Anyone who cannot adapt to the program will fail to continue the training because he/she has muscle injury, illness, pain, atrophy, increased cortisol and decreased physical performance. However, anyone who can adapt to the program will gain benefits from the training. These include hypertrophy of muscle fiber type I and IIa because it plays important role during the high intensity exercise for nearly 30 minutes (Table 1). This contributes to improve performance (Figure 4). Left ventricle muscle mass and chamber volume are increased. Mitochondria content (Figure 5), size and function, fat utilization during the exercise was increased leading to CHO sparing. Faster duration rates of oxygen diffusion and fuel into muscle. Oxidative enzyme level and efficiency and disposal of metabolic waste were also increased. Cell regulatory mechanism of metabolism was improved. Moreover, the exercise training increases insulin sensitivity via 3 possible mechanisms (Figure 6); 1) increased glucose transporter 4 exocytosis from its vesicle 2) decreased glucose transporter 4 endocytosis and 3) increased glucose transporter 4 activity.
Figure 1: Energy pathway (modified from refs.)

<table>
<thead>
<tr>
<th>Features</th>
<th>Type I fibers</th>
<th>Type II a fibers</th>
<th>Type II x fibers</th>
<th>Type II b fibers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Contraction time</td>
<td>Slow</td>
<td>Moderately Fast</td>
<td>Fast</td>
<td>Very fast</td>
</tr>
<tr>
<td>Resistance to fatigue</td>
<td>High</td>
<td>Fairly high</td>
<td>Moderate</td>
<td>Low</td>
</tr>
<tr>
<td>Activity Used for</td>
<td>Aerobic activity</td>
<td>Long-term anaerobic activity</td>
<td>Short-term anaerobic activity</td>
<td>Short-term anaerobic activity</td>
</tr>
<tr>
<td>Maximum duration of use</td>
<td>Hours</td>
<td>Less than 30 minutes</td>
<td>Less than 5 minutes</td>
<td>Less than 1 minute</td>
</tr>
<tr>
<td>Power produced</td>
<td>Low</td>
<td>Medium</td>
<td>High</td>
<td>Very high</td>
</tr>
<tr>
<td>Mitochondrial density</td>
<td>Very High</td>
<td>High</td>
<td>Medium</td>
<td>Very high</td>
</tr>
<tr>
<td>Capillary density</td>
<td>High</td>
<td>Intermediate</td>
<td>Low</td>
<td>Low</td>
</tr>
<tr>
<td>Oxidative capacity</td>
<td>High</td>
<td>High</td>
<td>Moderate</td>
<td>Low</td>
</tr>
<tr>
<td>Major storage fuel</td>
<td>ATP, Triglycerides</td>
<td>ATP, Creatine phosphate, glycogen</td>
<td>ATP, Creatine phosphate, glycogen (little)</td>
<td>ATP, Creatine phosphate</td>
</tr>
</tbody>
</table>
Figure 2 Substrate utilization in trained white subjects (modified from refs\textsuperscript{6-8}).

Figure 3 Substrate utilization in healthy sedentary Thai subjects (modified from ref\textsuperscript{10}).
Faster diffusion of oxygen and fuel into muscle

Increased expression of fatique-resistance of slow twitch muscle fibers

Increased mitochondrial function

Increased cardiac muscle mass

Increased left ventricular dilation and chamber volume

Increased disposal of metabolic waste

Increased stroke volume

Increase carbohydrate sparing

Increased oxidative enzyme level and efficiency

Improved cell regulatory mechanisms of metabolism

**Figure 4** Physiological changes from high-intensity exercise (modified from refs\textsuperscript{12,13})

**Figure 5** Influences of exercise bout and duration and intensity on muscle fiber content (modified from ref\textsuperscript{15})
In addition, “Focus T25” is a successful program because it also encourages nutrition program. This controls balanced energy intake and expenditure. Nutritional “Focus T25” Meal Plan and a “Focus T25” 5 Day Fast Track meal Plan really keep it simple on what to eat and when to eat it. This helps the exercisers successful in gaining body shape and health.

Precaution

If the exercisers do it wrong such as performing without enough warming up or preparation (dress, shoes and food intake) or too much effort at the first start they may have muscle pain, fatigue or injury. They have to be careful about the jumping part by not jumping too high. This may prevent them from continuing the exercise.

In addition, the exerciser should stop the exercise if they have symptoms as recommended by the American College of Sport Medicine (ACSM).

American College of Sport Medicine (ACSM) Indications for Termination of an Exercise Test

These indications for termination of an exercise test consists of 2 categories; absolute and relative indications.

Absolute Indications

1. Suspicion of a myocardial infarction or acute myocardial infarction (heart attack)
2. Onset of moderate-to-severe angina (chest pain)
3. Drop in systolic blood pressure (SBP) below standing resting pressure or drop in SBP with increasing workload accompanied by signs or symptoms
4. Signs of poor perfusion (circulation or blood flow), including pallor (pale appearance to the skin), cyanosis (bluish discoloration), or cold and clammy skin
5. Severe or unusual shortness of breath
6. CNS (central nervous system) symptoms
e.g., ataxia (failure of muscular coordination), vertigo (an illusion of dizziness, movement), visual or gait (pattern of walking or running) problems, confusion
7. Serious arrhythmias (abnormal heart rhythms)
e.g., second / third degree AV block, atrial fibrillation with fast ventricular response, increasing premature ventricular contractions or sustained ventricular tachycardia
8. Technical inability to monitor the ECG
9. Patient’s request (to stop)

Relative Indications
1. Any chest pain that is increasing
2. Physical or verbal manifestations of shortness of breath or severe fatigue
3. Wheezing
4. Leg cramps or intermittent claudication (grade 3 on a 4-point scale)
5. Hypertensive response (SBP >260 mm Hg; DBP >115 mm Hg)
6. Pronounced ECG changes from baseline
   It is >2 mm of horizontal or down sloping ST-segment depression, or
   >2 mm of ST-segment elevation (except in aVR)
7. Exercise-induced bundle branch block that cannot be distinguished from ventricular tachycardia
8. Less serious arrhythmias (abnormal heart rhythms) such as supraventricular tachycardia

Keys for success with “Focus T25”
1. If you are older than 35 years old you should receive physical examination by a doctor before starting “Focus T25” program.
2. Start with active warming up and finish with cool down by active stretching for at least 5 minutes.
3. Following the DVD at your own pace and even stop exercising if you feel cannot perform it. This is important for injury prevention.
4. If you have muscle pain during the first few days, you should first use cold pack to alleviate pain and followed by massaging together with hot pack.
5. Get some antioxidant or protein supplementation in the first 30 minutes to help with recovery.19-21

References
4. http://www.choosehealth.net/HealthSavingArticles/EnergyToBurn_PartI.html