Diastematomyelia: a case report

Penny Martin, DipPhy
Cashmere Physio Practice, Christchurch

ABSTRACT
This report alerts physiotherapists to possible variations in spinal pathology, in this case diastematomyelia and diplomyelia (Type I and Type 2 split cord malformations) which are rare spinal lesions. Their diagnosis requires spinal imaging by either CT myelography or Magnetic Resonance Imaging (MRI). Potentially severe neurological complications of paraparesis or paraplegia can be associated with diastematomyelia. This report discusses the diagnosis and management of a thirteen year old female who presented to physiotherapy complaining of low back pain (LBP), following an injury playing netball. This initial episode of LBP resolved during a course of three physiotherapy treatments. One year later the patient returned with a similar presentation. She reported a history of intermittent LBP and a feeling of weakness in either her right or left leg. Plain spinal X-rays revealed spina bifida occultus, and a subsequent MRI scan revealed Type 1 split cord malformation (diastematomyelia) at the midlumbar level, a tethering lesion that effectively pinions the spinal cord. The patient underwent neurosurgery to untether the spinal cord, and subsequently became pain-free and returned to sport. Martin P (2007): Diastematomyelia: a case report. New Zealand Journal of Physiotherapy 35(1) 24-27. Key words: diastematomyelia, split cord malformation, spinal cord tethering.

INTRODUCTION
Low back pain in adolescents is relatively common and is chronic or recurrent in up to a third of cases (Tiamela and Viljanen 1997). Congenital spinal defects that pose risks to the patient, or which contraindicate certain physiotherapy interventions, are rare. This report describes a case in which a potentially serious spinal defect was suspected by the treating physiotherapist, leading to the patient undergoing corrective neurosurgery.

HISTORY
An active adolescent girl (Patient G) complained of intermittent back pain after falling at netball. Back pain was severe at injury time, and she was unable to complete the game. She continued over the next 12 months to have bouts of pain during competitive sport, associated with weakness in either leg. Each time the pain settled quickly and she returned to sport within a few days. When the physiotherapist recognised this recurring pattern, medical specialist referral was initiated and scans revealed a tethered spinal cord in the lumbar region that had to be corrected surgically.

If a physiotherapist had manipulated this young girl’s spine as treatment for recurring low back pain the consequence may have been paraparesis or paraplegia. Because the condition of diastematomyelia is rare and intervention could be dangerous, this case is presented in detail with reference to common findings in tethered cord syndrome (Table 1) and characteristics of pain in spinal cord tethering (Table 2).

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<th>Table 1: Common findings in tethered cord syndrome</th>
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<tr>
<td>• Pain</td>
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<td>• Gait problems, often quite subtle</td>
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<td>• Progressive urinary incontinence; symptoms include urgency, frequency and enuresis</td>
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<td>• Progressive faecal incontinence including urgency, frequency and encopresis</td>
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<tr>
<td>• Any motor or sensory deficits in lower extremities</td>
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<td>• Altered sensation in genital regions and during sex</td>
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<td>• Impotence</td>
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<td>• Muscle weakness</td>
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<td>• Muscle atrophy</td>
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<td>• Hyporeflexia, especially any change in pre-existing signs</td>
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<td>• Scoliosis/lordosis</td>
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<td>• Foot deformities</td>
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<td>- Skin abnormalities – herald marks</td>
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<th>Table 2: Characteristics of pain in spinal cord tethering</th>
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<td>• Pain in lower back and legs exacerbated by physical activity, especially any which involves flexion and extension of the lumbosacral area</td>
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<td>• Leg pain is often in medial, lateral, anterior or posterior aspects of thighs or legs</td>
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<td>• Groin pain or pain in the genitofecal area is common</td>
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<td>• Pain is not less when lying supine (as opposed to disc disease)</td>
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<td>• Straight leg raising causes no difference to pain</td>
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<td>• In adults, pain governed by three “B” signs (usefulness will depend upon any pre-existing neurological and orthopaedic disabilities):</td>
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<td>- inability to sit with legs crossed (like Buddha)</td>
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<td>- difficulty in bending slightly at the waist with activities such as washing dishes</td>
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<td>- holding a baby or light material (&lt;2.5kg) at the waist level while standing</td>
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HISTORY
A thirteen year old girl presented without medical referral after an injury at netball. She complained of severe pain (visual analogue scale, 8) in her back and left buttock. After further games the pain reoccurred and was intermittent. Pain was worse when she was getting onto her bed and lying down in bed. She had no pre-existing serious back injuries and her general health was excellent. She was a Year ten student with no psychological history. She played competitive netball, tennis, horse riding, sailing, and skiing.

ON EXAMINATION
There was tenderness over the left paravertebral L2/3 level. Passive hip flexion at 130° caused back pain. Straight leg raise (SLR), left leg, caused pain at 80°. Back was painful on slump but not increased with knee extension or ankle dorsiflexion and not decreased with neck extension. Standing in extension with hip and knee flexion increased back pain. Tenderness was present over L4/5 and L5/S1 segments and the left lumbar paravertebral muscles. There were no neurological change; calves were of equal sizes. Bilaterally, the fifth toe was over-riding fourth toe (see Table 1). She had a small sacral dimple. An X-ray revealed spina bifida occulta L5.

ANALYSIS
Using Accident Compensation Corporation (ACC) terminology, there were no "red flags", meaning contraindications for physiotherapy treatment (A.C.C. guidelines 1999). The intermittent back and left buttock pain was restricting sporting activities. The reproduction of pain she had on lumbar extension with hip and knee flexion may be a sign of pars pathology. There was no night pain, but pain on getting into the lying position (See Table 2). There was tenderness over the paraspinal muscles. This adolescent girl was in good health. She had a small sacral dimple, which is common as most spina bifida patients have some form of cutaneous skin changes near the lumbosacral spine (for example dimple or scarred patch of skin shown in Table 1). As there was no association of underlying abnormalities in this case the sacral dimple had no relevance.

DIAGNOSIS
Non-specific low back pain.

TREATMENT
The diagnosis of non specific back pain was explained, with reassurance that back symptoms were common in adolescents and that back pain would diminish within three months (Taimela and Viljanen 1997). It was advised that netball should be discontinued until the symptoms were less irritable.

Pain relief measures used were:
A. Trial of soft tissue massage and trigger point therapy (Kovacs et al. 1997).
B. Ultrasound therapy: Frequency of One MHz (millions of cycles per second) at intensity pulsed 0.5 W/cm² for six minutes over left mid and low lumbar paravertebral muscles. A pro-inflammatory effect was expected (Dyson 1987, Maxwell 1992, Nussbaum et al. 1996, Nussbaum 1997).
D. Mobilisation. Mulligan double leg rotation to the right (patient lies supine with hip and knees flexed and physiotherapist rotates legs within pain free range to the side of the limited SLR).

OUTCOME AND PROGRESS
Pain levels rapidly improved during the three initial physiotherapy treatments in the first week. However, straight leg raise signs remained unchanged. Patient G returned to competitive netball and did not return to the physiotherapy clinic.

One year later Patient G attended the physiotherapy clinic complaining of similar symptoms following reinjury at netball. She complained of occasional back pain with associated left leg weakness. The on-court physiotherapist had treated her with acupuncture which she felt eased symptoms (Ernst et al. 1998). An X ray was taken on 5/7/2. The result read, “Spina bifida occulta (of no concern)”. This girl returned to sport: the symptoms of back and buttock pain resolved spontaneously. Occasionally she would have an awareness of back pain and a feeling of weakness in either left or right leg.

REFERRAL TO MEDICAL SPECIALIST
Reasons for this referral included the age of patient, recurring episodes, the spina bifida occulta x ray finding: symptoms of leg weakness varying from left to right (subjective, not reproducible on examination), and pressure from the netball coach.

An MRI scan of the thoracolumbar spine revealed a congenital abnormality in the mid lumbar region. There was a very thin syrinx cavity (terminal syringomyelia) within the lower/caudal spinal cord extending from T12 to L2. The spinal cord then became bifid at L1 down to L3 with the two hemicords being of equal size. The conus was low being located at the L4 level where it continued caudally as a thickened filum terminale. There were two dural tubes over a short 5mm length at the L3 level, each containing the equal-sized hemicords. At L3 there was a very thin bony/ fibrous spur extending antero-posteriorly across the spinal canal, connecting the posterior body of L3 to anterior surface of the L3 laminae/neural arch. These appearances were those of a Type 1 Split Cord Malformation, or diastematomyelia.
THE SPLIT CORD CONDITION

Split Cord Malformations (SCM) (Pang 1992) are classified as Type 1 (two dural tubes — diastematomyelia) and Type 2 (one dural tube — diplomyelia). It is a complex defect which occurs during the 4th week of foetal development. SCM is a rare form of spinal dysraphism which occurs as a result of the foetus having an accessory neurorneric canal. This canal lies in the midline and therefore the developing spinal elements (vertebrae, dural tube, spinal cord and spinal nerves) are separated (split) and develop as two sets of structures. These are separated by the mesenchymal components in the midline, being combinations of bone (the spur), cartilage and fibrous tissue. There is always a sagittal cleft of varying extent, with a “split” spinal cord, and the hemicords are more often of equal size but may be asymmetrical with musculature on the leg on the side of the smaller hemicord being typically of lesser diameter. The cords re-unite a variable distance cephalad to the spur or tethering lesion, and it is here that the problem of tethering occurs. In Type 1 SCM each of the two dural sacs is occupied by a hemicord, and the residual mesenchymal elements include a bony spur which lies between the dural tubes. It is this rigid spur that prevents normal spinal cord mobility, and movement with growth and physical activity will usually render the patient symptomatic. In Type 2 the tethering lesion is a thick fibrous band (called a myelomeningocele manqué) which attaches to one or both of the hemicords and also to the dura. SCMs occur most commonly at mid to high lumbar levels, less commonly at low to mid thoracic levels, and least commonly in the cervical spine.

PATIENTS WITH SCM PRESENT IN 4 WAYS:
1. With a visible abnormality in the skin lesion or lumbosacral region: lesions include a hairy patch (hypertrichosis); dermal sinus; midline pigmented area; asymmetry of the buttocks or gluteal fold; asymmetry of the leg musculature —typically of the calf muscles.
2. Pain/neurological syndrome: back pain and/or leg pain after exercise especially if associated with intermittent leg symptoms of weakness, gait disturbance; tingling, numbness; progressive weakness or sensory changes in the legs; sphincter disturbance (bladder in particular); progressive scoliosis, sympathetic dystrophy.

3. In patients with known spina bifida (present or past meningocoele or myelomeningocele) who develop a neurological deficit/sensory level that is above that attributed to the original spina bifida lesion.

4. There have been case reports of patients with SCM sustaining significant or severe spinal cord damage following minor injury, including falling over and also after spinal manipulations.

**SURGERY**

A neurosurgical operation was performed on the 22nd July 2003.

The un tethering of the spinal cord was achieved by:

1. Excising the bony spur which ran between the two dural tubes — it extended all the way across the spinal canal from posterior to anterior and the anterior third of the spur was cartilaginous. The bony spur took up sixty per cent of anteroposterior width of canal and became cartilaginous over the anterior 3mm.

2. Converting the two dural tubes to a single dural tube.

3. Sectioning the thickened filum terminale.

**OUTCOME**

This adolescent girl remained neurologically intact following surgery and is now pain free, fit and active, and has returned to competitive sport.

**CONCLUSION**

Not all positive SLR signs are reversible by manual neural tension or physiotherapy techniques, and manipulation could have disastrous effects including paralysis of lower limbs or bladder/bowel incontinence. This condition, if left untreated or undiagnosed, could have deleterious consequences in terms of impaired spinal cord function (Pang 1992). Although uncommon (only 11 operations for SCM have been undertaken in Christchurch in the last 22 years), an MRI is necessary to rule out this pathology.

The ACC has released guidelines for physiotherapists purporting to describe evidence based treatment. The guidelines advise that non specific low back pain can be managed by manipulation, exercise and education. The guidelines do not encourage referral for X-ray. This case demonstrates the necessity for physiotherapists to take a careful history and carry out a thorough examination. Continuing observation during a course of treatment is essential with cross-referrals within the health team being made when it is deemed necessary. Conservative treatment during a period of observation was justified in this case and further damage, which could have been severe, was avoided.

**ACKNOWLEDGEMENTS**

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**ADDRESS FOR CORRESPONDENCE:**

Cashmere Physio Practice, 4 Colombo Street, Cashmere, Christchurch.