Relationship Between Sacral Dimples and Spinal Cord Malformations in Children

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Abstract

Background: Traditional teaching states that if the base of a sacral dimple is visible, associated spinal cord abnormalities are unlikely. In our experience, we noted that in several patients this was not true. We therefore set out to evaluate the incidence of spinal cord abnormalities in a cohort of children with sacral dimple.

Objective: Our objective was to determine the frequency of spinal cord abnormalities detected on whole-spine MRI in children with pure sacral dimple (cutaneous dimple without associated local pigmentation or hairy patch). The frequency of neurosurgical intervention in those with a spinal cord malformation was also calculated.

Materials and methods: We retrospectively reviewed all patients under the age of 16 years with clinically confirmed pure sacral dimple, who had undergone spinal MRI between 2005 and 2016. Patients with coccygeal pits were excluded from the study. We analyzed the MRI findings, calculated the incidence of cord abnormalities, and proportion of those who subsequently underwent neurosurgical procedures, such as spinal cord untethering. Other associated abnormalities were documented.

Results: During the 11-year study period, 33 patients (19 boys) with confirmed sacral dimple and whole-spine MRI were identified. The median age at the time of MRI was 21 months (range 2 months to 13 years). Eleven patients (33.3%) had abnormal cord detected on the MRI. Out of them, 4 had tethered cord, 2 had low lying cord (L2/3 level). Five had lesions related to the filum terminale (dermoid, cyst, lipoma). Of the four patients with tethered spinal cord, 2 had untethering procedures, 2 so far have been managed conservatively. Of the 7 patients with other cord abnormalities, 1 had excision of sacral dermoid. Nine of the 33 patients (27%) had other congenital anomalies, including 5 with syndromes.

Conclusion: In our experience, the incidence of spinal cord abnormalities in children with pure sacral dimple was 33.3%, some of which required surgery. Other congenital abnormalities were common. We recommend whole spine MRI and a careful search for other congenital abnormalities.

Keywords: sacral dimples, spinal cord malformations, coccygeal pits, tethered spinal cord

Introduction

A simple sacral dimple in children is defined as a dimple located within 2.5 cm from the anus, superficial, and less than 0.5 cm in diameter – with no additional associated cutaneous markers, such as local pigmentation or a hairy patch in children [1, 2].

Sacral dimples are common, occurring in up to 3-8% of the population [3]. The majority of these dimples are minor and do not represent any underlying disease [3]. However, they can manifest with serious and long term neurological, urological and cutaneous abnormalities [4]. They are usually detected in post-natal checks by a

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paediatrician, and the following features are assessed.

- Is the base of the dimple visible? If not, this may indicate that the neural tube is not completely closed.
- Is there a tuft of hair in the dimple? This may also indicate a neurological abnormality.
- How close to the buttocks is the dimple? The lower it is, the lower the risk of neurological abnormalities.
- Is the lower limb neurology, bladder and bowel function normal?
- Are there any other additional features of a congenital syndrome? Examples include Miller-Dieker and Smith-Lemli-Opitz syndrome [5].

Sacral dimples are the most commonly seen cutaneous anomalies on paediatric spinal examination [5]. The more obvious lesions may be a manifestation of underlying tethered cord, justifying spinal cord imaging [1][2], such as spinal MRI scan [5], or in younger babies, spinal ultrasound initially.

However, given that the presence of a simple sacral dimple in an otherwise healthy child is not always associated with underlying spinal cord tethering [1], there have been studies suggesting that further evaluation with imaging is not necessary [2]. In fact, there is no clear guideline on an appropriate diagnostic work-up in the small subset of patients with pure sacral dimple and no overlying skin changes [1].

In our experience, we have found that many of our patients with a pure sacral dimple do have associated spinal cord abnormalities. Thus, the aim of this study was to evaluate the incidence of spinal cord abnormalities in children with a pure sacral dimple.

**Material and Methods**

We performed a retrospective study of all children with a clinically confirmed simple sacral dimple who underwent spinal MRI at Queens Medical Centre, Nottingham, between 2005 and 2016.

We requested the list of patients who had MRI of the spine to assess the sacral dimple in children under the age of 16, performed in the last 11 years. Patients with coccygeal pits and children in whom the dimples had overlying pigmentation or hairy patches were excluded from the study.

Details of all patients included were then accessed from our patient database. We collected data on patient demographics, presenting symptoms, and features of the dimple but also associated abnormalities, particularly of neurological, urological or bowel-related problems, in addition to making note of whether there were any clinical features suggestive of associated underlying syndromes (including whether these were later confirmed). Past medical and surgical history was also recorded.

Imaging performed in addition to the MRI was also recorded. In most cases, this included ultrasound (US) scans performed prior to referral, and a MRI of the brain in a few patients. The findings of the spine MRI were recorded, followed by indication for further management: “reassured and discharged” if MRI was normal, “conservative with regular follow up” if an abnormality was present where surgery was not required at that time or “surgical intervention”, for significant cord pathology.

We then calculated the incidence of spinal cord abnormalities within the 11-year period, and the proportion of these patients who subsequently underwent neurosurgical intervention.

**Results**

During the 11-years study period, 33 patients under the age of 16 underwent whole-spine MRI to further assess a simple sacral dimple. Of the 33 patients, 19 were male (57.5%) and 14 were female (42.4%). The age of the patients ranged from 2 months to 13 years with a median of 21 months. Five patients (15%) had associated syndromes, including Cornelia de Lange, Keratitis-ichthyosis-deafness (KID) syndrome, Aicardi-Goutières, caudal regression syndrome, one unknown syndrome. Nine patients (27%) had neurological abnormalities such as hypotonia, global developmental delay and gross motor problems. Thirteen patients (33%) had general urological problems such as a duplex kidney, hypospadias, urine incontinence or a bladder diverticulum. Three patients (9%) had cardiac abnormalities, such as tetratology of fallot (TOF), ventricular septal defect (VSD) or atrial septal defect (ASD) (Fig. 1).

In terms of previous surgical history, of the 33 patients, 1 had a duodenal atresia repaired, 2 had undergone pyloromyotomy, another 2 had major cardiac surgery (TOF, VSD and ASD repair, pulmonary valvotomy), 2 underwent hypospadias repair and 1 underwent a percutaneous nephrolithotomy (PCNL) of the right kidney.

Seven patients had a spinal ultrasound performed prior to being referred. Five of these were normal while 2 required a spinal MRI to further assess a low-lying cord. Three patients also had a renal ultrasound performed (all normal) and 8 also had brain MRI. Of the latter, 1
showed ventricular enlargement, and one showed mild prominence of extra cerebral CSF space. The remaining 6 patients had a normal MRI of the brain.

In terms of neurological abnormalities, 11 patients (33.3%) had an abnormal cord detected on the whole spine MRI. Of the 11 patients, 4 (12.1%) had a tethered cord, 2 (6.06%) had a low-lying cord (L2/3 level) and 5 (15.2%) had lesions related to the filum terminale (dermoid, cyst, lipoma).

Of the four patients with a tethered spinal cord, 2 underwent untethering procedures for symptoms, whilst the remaining 2 are currently being managed conservatively with regular follow up. Of the remaining 7 patients with other cord abnormalities (low lying cord, filum terminale lesions), 1 underwent an excision of a sacral dermoid. All of these 7 patients had a good outcome (Fig. 2).

It was noted that of the 11 patients with an abnormal cord found on MRI, 3 had associated syndrome (Cornelia de Lange, Keratitis-ichthyosis-deafness (KID) syndrome, Aicardi-Goutières) and there was a relatively higher incidence of urological related problems.

In summary, of the 33 patients with a simple sacral dimple, 9 (27%) had congenital anomalies, 11 (33%) had spinal cord abnormalities (including 3 syndromic children) with only 4 (12%) found to have a tethered cord.

**Discussions**

Previously published studies have suggested that the risk of spinal malformations in asympto-
motic, healthy infants with an isolated simple sacral dimple is exceedingly low [1]. Alongside this, several studies have suggested that there is little value in investigating a simple sacral dimple with further imaging [5], whilst others suggest simply performing a screening ultrasound scan [1]. Importantly, there is no clear guidance on an appropriate diagnostic workup in this group of children [1]. Nevertheless, through the 11-year period of our study, we have demonstrated a high yield (33.3%) of spinal cord abnormalities on a MRI of the spine, emphasizing the importance of imaging in patients with a simple sacral dimple. This has not been demonstrated in previous studies.

Kucera et al. [1] performed a 12-year study in children with a simple sacral dimple in USA. From a large cohort of asymptomatic children, only 2.1% of children were found to have spinal cord abnormalities on a spinal US or MRI scan. Out of 3,884 children, only 128 (3.3%) had an abnormal US of the spine, with 76 (2.1%) children found to have a spinal abnormality on follow up imaging. In this study, the large patient selection would allow the results of this study to be easily applied to other practices. Although our data was collected over an 11-year period, the number of patients who had a MRI of the spine to investigate a simple sacral dimple was only 33, which may be an important limitation in our study.

Although the Kucera et al. study [1] had a large patient sample, this consisted of patients who had a screening ultrasound of the spinal cord performed for a simple sacral dimple. A follow up MRI or US was then reviewed in all those with an abnormal screening US scan. However, this would exclude several patients who had a US reported as normal, although further imaging may have shown an abnormality. Our inclusion criteria consisted of all patients that had an MRI spine requested to investigate a simple sacral dimple. Within this group, a small proportion of patients had an US performed prior to referral. We found that not all of the US findings correlated with the subsequent MRI findings. In fact, at least one patient had an US scan reported as normal, only to have a follow up MRI scan showing tethering and a cutaneous sinus, requiring surgical un-tethering. Thus, this further emphasizes the importance of requesting an MRI of the spine for all patients with a simple sacral dimple, as an US scan alone can be inadequate. This may be due to the fact that an US is user-dependent, and requires an experienced sonographer.

Sonographic examination of the paediatric spinal canal is accomplished by scanning through the normally incompletely ossified posterior elements of spine. It is most successful in the new born period and in early infancy. Infants older than 6 months, the examination is very limited [6]. Another limitation is the retrospective nature of our study. Since the method of collecting data was to include all whole spinal MRI scans investigating a simple sacral dimple, this would not have included patients who may have been identified as having a simple sacral dimple, but either not referred, were investigated with an US scan with no follow up MRI, or had the MRI performed elsewhere. Thus, our patient selection could under-represent actual data. Our center is a tertiary center and we receive referrals from all over our region. Patients with sacral dimples who complains of symptoms like wetting, incontinence, urgency or urinary tract infections are referred to us for review and further investigations. They are reviewed in our urological department and we request spinal MRI scans to rule out any pathology. If there is any associated and related pathology we referred it to the neuro-surgical department for their opinion and review. All the patients referred to our urological department or neurosurgical department had a spinal cord imaging. As we are a tertiary center not all patients within our catchment area are referred to us, as they are managed locally.

Another recent study addresses concerns regarding investigating a simple sacral dimple for underlying spinal cord malformations with ultrasound. Albert GW [7] has looked into nine papers where 3.4% of 5,166 patients had an abnormal USS finding, although most of these findings were of no clinical significance. However, given the fact that a missed tethered cord in a patient with a simple sacral dimple can lead to significant neurological dysfunction, orthopaedic deformities, as well as genitourinary and gastrointestinal abnormalities [4], we believe that the low percentage of patients that require surgical intervention as shown in previous studies should not prevent further investigation in all patients with a simple sacral dimple. Of the 33 patients, 12% were found to have a tethered cord. The percentage of children with a tethered cord in our dataset is exceptionally high, and this further supports our department protocol to obtain further imaging in patients with a simple sacral dimple. In our tertiary center, a selected group of patients are referred from primary care or district centers. These patients are of concern or are having urological problems. Initially they will have been reviewed by the local GP or a
pediatrician, and managed at that level. Depending upon the initial examination and symptoms they are then referred to our urological department for review and further management. Only patients with problems are referred to our urological or neurosurgical department. They all have had a MRI of the spine as they were difficult to treat or having ongoing problems. This study has provided new information, and at present, will support our departmental guidelines in terms of organizing a whole-spine MRI scan for all children with a pure sacral dimple. Further research is required with a larger sample of patients, ideally in a prospective study, in order to provide results that can be applied nationally.

Conclusion
In our experience, the incidence of spinal cord abnormalities in children with a simple sacral dimple was 33.3%, with just over a third (12%) with a tethered cord. Surgery was only required in 3 (9.09%) patients. Other congenital abnormalities were also common in this set of patients. As a result, we recommend that all patients with a simple sacral dimple should undergo whole-spine MRI. There should also be a careful search for other congenital abnormalities, as these were common in our study group.

REFERENCES