CONGENITAL DERMAL SINUSES

BACKGROUND. Congenital dermal sinuses represent cutaneous depressions or tracts that are lined by stratified squamous epithelium. They communicate between the surface of the skin and deeper structures and may occur anywhere along the craniospinal axis. These sinuses are thought to result from abnormal separation of the cutaneous and neural ectoderm between the third and fifth week of intrauterine life. They may be often accompanied by other cutaneous stigmata, various dysraphic abnormalities, or intraspinal tumors.

In the sacrococcygeal area, cutaneous congenital abnormalities are relatively common. It is estimated that 2% to 4% of children harbor intergluteal dorsal dermal sinuses. These intergluteal sinuses in the perianal region are frequently referred to as pits or dimples. Their cause is considered similar to other congenital dermal sinuses and appears unrelated to acquired pilonidal conditions observed in adults. They may become susceptible to local recurrent infection from trauma or hirsutism.

Controversy regarding the evaluation and management of cutaneous defects in the coccygeal region exists.

METHODS. Both a literature review and a career review of clinical material were performed. Databases for articles published in English were surveyed for key words related to coccygeal sinuses using standard computerized search techniques. The medical records of children presenting to our neurosurgical clinic for evaluation of dorsal dermal sinuses were reviewed to identify those with intergluteal sinuses.

RESULTS. In the evaluation of reported cases and of our own, we were unable to identify any children with coccygeal sinuses without other cutaneous markers other than hair with findings suggestive of intraspinal communication.

CONCLUSIONS. Intergluteal dorsal dermal sinuses are relatively common lesions that frequently come to neurosurgical attention. They do not seem to be associated with significant risk of spinal cord and intraspinal abnormalities. Simple intergluteal dorsal dermal sinuses without other cutaneous findings do not require radiographic or surgical evaluation and treatment. If other markers or neurologic symptoms are present, however, radiographic evaluation may be indicated. Pediatrics 2000;105(5). URL: http://www.pediatrics.org/cgi/content/full/105/5/69; occult spinal dysraphism, spina bifida occulta, dermal sinus, pilonidal sinus.

REFERENCES

ABBREVIATION. OSD, occult spinal dysraphism.
sinuses that occur below the natal cleft in the peri-
anal region are frequently referred to as pits or dim-
ples. They may become susceptible to local recurrent
infection from trauma or hirsuitism. They are not
related to acquired pilonidal conditions observed in
adults.12 Their cause is not entirely understood.

Controversy regarding an association between
coccygeal pits and spina bifida or any communica-
tion with the subarachnoid space exists in the litera-
ture. Some authors argue that the presence of any
cutaneous abnormality in the gluteal region warrants
radiographic and/or surgical evaluation because of a
suspected association with abnormal communica-
tions with or various abnormalities of the contents of
the intraspinal cavity.13,15–18 Some clinicians suggest
that the respective appearance of the lesion should
determine its further work-up. Lesions are inspected
for the ability to discern the cutaneous base or for the
presence of hair.13 Others, however, believe that all
coccygeal dimples or sinuses are innocent and war-
rant no additional evaluation other than physical
examination.19–22 Hence, the proper evaluation and
management of these isolated cutaneous defects in
the coccygeal region are relatively uncertain.

Given the common occurrence of these cutaneous
abnormalities in children, any statement requiring
diagnostic evaluation and/or surgical exploration
for all coccygeal pits is of a public health concern.
Do patients with coccygeal pits warrant investigation or
treatment for possible intraspinal anomalies or infec-
tion? In an attempt to determine the appropriate
therapeutic assessment of these intergluteal abnor-
malities, 2 tasks were performed.

METHODS

We searched the medical literature for published studies con-
cerning the association between coccygeal pits and spinal dysra-
phism and/or infection. Using standard computer search tech-
niques, articles written in English containing the following key
words were reviewed: dermal sinus, pilonidal sinus, spina bifida
oculta, OSD, congenital dermal sinus, and sacrococcygeal dermal
sinus. Original and review abstracts and articles were evaluated.
The bibliographies of the relevant articles were examined to iden-
tify additional studies of association. The 2 investigators reviewed
all reports.
In addition, the medical records of all children presenting to
our neurosurgical clinic for evaluation of dorsal dermal sinuses
between July 1978 and July 1998 were reviewed. The clinic
spanned 2 academic institutions during the study. The clinical
presentation, radiographic evaluation, and subsequent manage-
ment of patients were studied to identify appropriate individuals
for inclusion. The clinical evaluation consisted of a detailed neu-
rological and general physical examination in all patients.

A uniform definition was applied to the diagnosis of an isolated
coccygeal pit: a cutaneous pit, dimple, or sinus located below the
level of a symmetric intergluteal crease that is without the asso-
ciated presence of any additional cutaneous anomaly. Children
were excluded from additional review if hemangiomas, abnormal
tufts of hair, areas of cutaneous hypo- or hyper-pigmentation,
sinuses, dimples, or subcutaneous masses were identified any-
where on the back in addition to the presumed coccygeal lesion
(Fig 2). Children were also excluded by the presence of an asym-
metric gluteal cleft.

RESULTS

After an extensive and critical review of the En-
GLISH literature, only 7 cases of cutaneous, coccygeal
abnormalities associated with abnormalities of or ab-
normal communications with intraspinal contents
were identified (Table 1). These 7 individuals formed
the basis of 5 reports.13,15,24–26 Their clinical presenta-
tion varied. Six individuals presented with a neuro-
logic infection, bacterial meningitis affected 5, and a
spinal epidural abscess occurred in another. The final
patient was neurologically normal and without his-
tory of antecedent infection but underwent prophyl-
lactic surgical exploration. An intradural dermoid
tumor was identified.

Our literature review suggests that the relative risk
of associated neurologic infection or deficit is exceed-
ingly rare. Only 7 individuals have been reported in
the English literature to exhibit findings suggestive
of coccygeal pit in association with an intraspinal
abnormality or neurologic infection. Careful inspec-
tion of these published reports may reduce this small
number even further. In 5 cases, the coccygeal abnor-
mality was not in isolation.13,15,24 Additional dimples
and/or sinuses above the intergluteal crease and
hemangiomas were documented. The risk of associ-
ated OSD and neurologic infection has been clearly
demonstrated for such cutaneous abnormalities. The
presence of coccygeal pit, shown to be quite com-
mon, may have been incidentally present in these
patients. The serendipitous presence of the coccygeal
anomaly may have had nothing to do with the asso-
ciated neurologic abnormality.

Similarly, the description of exact location is incon-
clusive in the reports of 2 additional patients.25,26 The
terminology used for location description is in-
consistent and photographic documentation is lacking
with these respective reports. The sinuses described
in the reports by Ripley and Thompson25 and by Stammers26
may actually be located above the natal
cleft of the buttocks representative of well-character-
zized cutaneous signatures of OSD.

After a comprehensive review of the medical
records of individuals evaluated in our neurosurgical
clinic during a 20-year interval and exclusion of
those who exhibited additional cutaneous abnor-
malities, a total of 1000 patients with simple coccygeal
pits were identified. Nearly all patients were below 6
months of age. Evaluation was limited to clinical
examination and history. Radiographic imaging
studies were not routinely obtained unless per-
formed before referral. The patient ages ranged between 1 week and 20 years. No patient was found to exhibit any history of neurologic infection or neurologic deficit on either their initial evaluation or follow-up.

DISCUSSION

The general terms spina bifida and spinal dysraphism refer to those malformations involving any or all the tissues on the midline of the back. They are used to designate those spinal anomalies that possess an incomplete or an inadequate fusion of dorsal midline structures of the developing embryo. They represent a spectrum of deformities that include abnormalities of the skin, vertebral column, meninges, or neural elements that may occur alone or in combination. The extent of the malformations may be of a mild, moderate, or severe degree. Vertebral column abnormalities are invariably present with involvement of the spinal cord and meninges. Abnormalities of the skin are also common in such instances. Hence, the detection of a subtle cutaneous anomaly in a child may be crucial to future neurologic, urologic, and orthopedic development.

OSD refers to lesions that are concealed without exposure of neural tissue or cystic masses. The location and nature of the neural malformation is less obvious on physical examination than overt forms of open spina bifida. They are a heterogeneous group of conditions that are categorized together because of their common embryological origin and the tendency for multiple pathologic entities to be expressed simultaneously in a single individual. Examples include the tight filum terminale, intraspinal lipoma, split cord malformation, dural sinus and inclusion tumor/cyst, neurenteric cyst, meningocele manque, and myelocystocele. The exact incidence of OSD in the general population is not entirely clear. Many defects remain undiscovered and persist without evidence suggestive of neurologic, musculoskeletal, or urologic impairment into adult life. These occult forms of spinal bifida are much more common than are those that are open. The natural history of OSD is variable and often unpredictable. Although some individuals remain asymptomatic throughout adulthood, others may develop progressive dysfunction of the lower limbs and bladder. The insidious fashion in which such complications develop may lead to irreversible damage before any symptomatic manifestation. The risk of neurologic deterioration exists at all ages. It increases with time and is frequently progressive. Neurosurgical intervention has been demonstrated to halt progression of neurologic deficits.

The optimal management for the multiple abnormalities of OSD includes early diagnosis, neurosurgical referral, and surgical intervention. The primary problem with these conditions is not the risk of intervention, but actually the identification of which individuals are at risk for neurologic compromise and the recognition of the earliest possible clinical manifestations that will provide their detection. Clinical abnormalities may vary according to age. They may bear no obvious relationship to the nervous system. In addition, monitoring the bowel and bladder function in a young child is difficult and too often postponed until an age consistent with urinary continence is reached and irreversible deficits are already present.

Cutaneous signatures are often the initial marker of congenital spine abnormalities and are the most common finding leading to investigation. It is estimated that over one half of individuals with OSD exhibit such stigmata at presentation. They tend to occur in the midline of the back and are often located at the level of the intraspinal abnormality. They are most commonly identified in the lumbarosacral region. Numerous cutaneous lesions have been described that may occur singularly or in combination. Superficial lesions include areas of abnormal or unusual patterns of hair growth, hemangiomas, paraspinal telangiectasias, areas of hyper- or hypopigmentation, lobulated fatty subcutaneous masses, skin tags or tails, asymmetrical gluteal creases, and dermal sinuses or dimples.

Congenital dermal sinuses are cutaneous depressions or tracts that are lined by stratified squamous epithelium. They can signify both the occult dysraphic state and the presence of a connection between the skin surface and subarachnoid space. They may be difficult to identify and can be located anywhere along the craniospinal axis. They are thought to develop in response to an abnormal separation of the cutaneous and the neural ectoderm between the third and fifth weeks of intrauterine life. They are frequently associated with other cutaneous abnormalities, various dysraphic lesions, or intraspinal tumors.

In the sacrococcygeal region, cutaneous congenital abnormalities are common. In a prospective search for congenital dermal abnormalities of the craniospinal axis, Powell et al examined 1997 consecutive newborns delivered at a single institution during a 1-year period. Approximately 3% of the neonates exhibited significant paraspinal abnormalities above the intergluteal crease, while 4.3% of children exhibited coccygeal pits. Hence, these intergluteal abnormalities are not infrequent.
They may become susceptible to local recurrent infection from trauma or hirsuitism. They are not related to acquired pilonidal conditions observed in adults.12 Their cause is not clear. Controversy regarding an association with OSD and the proper evaluation and management of isolated cutaneous defects in the coccygeal region exists.13,15–23 Given the relative frequency of these cutaneous abnormalities, any statement requiring diagnostic evaluation is of public health concern. Based on our studies, it becomes difficult to recommend surgical treatment or even radiographic evaluation for isolated coccygeal pits.

Retrospective review of our own patient data supports the innocence of coccygeal pits. Although such data can be criticized for lacking radiographic documentation, others have already demonstrated evidence of radiographic benignity. Herman et al35 performed spinal ultrasound on 53 infants with coccygeal pits. The average age of those studied was 24 days. The location of the conus medullaris was found to be between T12 and L1 in 13%, behind the L1 vertebral body in 20%, and behind the L2 vertebral body in 67%. No intraspinal anomalies were identified.35 Gibson et al36 prospectively examined 95 neonates harboring cutaneous abnormalities of the back with ultrasound. Seventy-five of the 95 children had isolated coccygeal pits. No abnormality of the spinal axis was identified in those with coccygeal pits.36 The radiographic data appear to correlate and follow our clinical impressions that isolated coccygeal pits are benign. Hence, the burden of proof is not with us to radiographically demonstrate that simple

Fig 2. Photographs of the low back and buttocks of infants excluded from this study. All children possessed a coccygeal pit in addition to a variety of cutaneous signatures for OSD. A, cutaneous hemangioma; B, abnormal tuft of hair; C, an additional dermal sinus, above the intergluteal crease; and D, cutaneous hemangioma and subcutaneous lobulated fatty mass.
Coccygeal pits are benign. The burden of proof must rest with those who mandate radiographic or even surgical investigation. To this, there seems to be no justification.

CONCLUSIONS

Coccygeal pits are very common abnormalities of the skin. Lesions in isolation are associated with a small incidence for associated neurologic infection or neurologic deterioration. Therapeutic evaluation may be limited to physical examination. Lesions in association with other well-defined cutaneous stigmata of OSD warrant further radiographic and/or surgical inspection.

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