

# Congenital Dermal Sinuses : An Clinical Analysis of 20 Cases

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**Objective :** Congenital dermal sinus is a rare congenital disease that results from the failure of the neuroectoderm to separate from the surface ectoderm during the process of neurulation, where there is communication between the skin and the deeper structures. Their pathogenesis, clinical course and treatment strategy are well known. We analyze our series and compare our results with other series.

**Methods :** Twenty patients were diagnosed as congenital dermal sinus and confirmed pathologically from October 1986 to July 2003 at our hospital. We studied the patients' clinical manifestations, radiological findings and pathological profiles.

**Results :** Seven cases were located in the suboccipital area and 13 cases were located in the spinal area. Interestingly, 4 of 13 spinal lesion cutaneous openings were located lower than the 3rd sacral body level. 8 of 20 lesions were terminated at neural structures, 4 of 20 lesions were terminated at the intradural portion and others terminated at the extradural portion. Nine anomalies were combined with the dermal sinus, including 4 lipomas, 2 Currarino's triad, 1 encephalocele, 1 myelomeningocele and 1 diastematomyelia. Eleven patients had dermoid tumors.

**Conclusion :** Congenital Dermal Sinus must be surgically removed immediately if they are diagnosed. The surgical procedure of congenital dermal sinus is complete removal, but in some cases, complete removal is impossible. In those cases, we removed all epithelial tissues. We consider sacrococcygeal dimple almost invariably have no connection with intraspinal structures. But, if other cutaneous manifestations are combined with cutaneous pits, it can communicate with the sacrococcygeal dimple.

**KEY WORDS :** Congenital dermal sinus · Squamous epithelium · Surgery.

## Introduction

Congenital dermal sinuses (CDS) are well known diseases. Its pathogenesis, clinical features, diagnostic tools and treatments are also well established. But its prevalence is not so high that individual clinicians may document many case studies. That's why diagnostic mistakes may happen in some clinics. We analyze our experiences over 17 years and compare them with other series.

## Materials and Methods

### Materials

Twenty patients were diagnosed as CDS and confirmed pathologically from October 1986 to July 2003 at our hos-

pital (Table 1). We checked the patients' clinical manifestations, radiological findings and pathological profiles. At the time of the operation, the patient's ages were from 2 months to 12.8 years. The median age was 2.2 years, and the mean age was 3.6 years. Only patients under the age of 15 years were studied at the children's hospital. Exclusion criteria were : ① not infected solitary lesions below the 3rd sacrum level, ② caudal directed sinuses, ③ sinuses that were not penetrating beneath subcutaneous tissue, and ④ sinus walls not covered with squamous epithelium.

### Methods

We reviewed the patients' medical records and radiographs, especially, the patients' chief complaints, skin manifestations, nervous system infections, neurological signs, radiographic findings, combined anomalies, combined tumors and postoperative courses.

1) When CDS was diagnosed, clinicians thought that it should be operated on as soon as possible<sup>2,5)</sup>. We want to verify this concept through our series.

2) We checked the patients' clinical manifestations on

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**Table 1.** Summary of the cases

Name	Location	Cut. Opening	Termination	Asso. Anomaly	Combined tumor	Operation
Case1	LS	sacrococcyx	paraspinal m.	LMMC	none	complete
Case2	Occipital	suboccipital	subarachnoid	none	dermoid	complete
Case3	Occipital	suboccipital	neural element : Cbil.	none	dermoid	complete
Case4	T	T10-11	neural element : cord	none	dermoid	complete : coagulation
Case5	Occipital	suboccipital	extradural	encephalocele	dermoid	complete
Case6	T	T4	neural element : cord	MMC	dermoid	complete : coagulation
Case7	LS	sacrococcyx	paraspinal m.	Currarino, LMMC		complete
Case8	LS	below S3	neural element : conus amputated at	none	dermoid	complete incomplete : common wall
Case9	Occipital	suboccipital	extradural portion	none	none	with sinus
Case10	Occipital	C1/2	neural element : cord	none	dermoid	complete
Case11	Occipital	suboccipital	extradural	none	dermoid	complete
Case12	LS	dual opening(Lt. Butt & midline)	subarachnoid : Butttock, extradural : midline	LMMC	none	complete
Case13	LS	dual opening(Lt. Butt X2)	subarachnoid : intradural lipoma	LMMC	none	complete
Case14	CT	C7/T1	extradural	SCM type II	none	complete
Case15	LS	L5/S1	neural element : rootlet	none	none	complete
Case16	Occipital	suboccipital	paraspinal m.	none	none	complete
Case17	LS	3cm above anus	neural element : cord	LMMC	none	complete
Case18	LS	L5	subarachnoid	none	dermoid	complete
Case19	LS	dual opening(bilat.S1/2)	paraspinal m.	Currarino	dermoid (X2)	complete
Case20	TLS	S2	neural element : cord	none	dermoid	incomplete : multiple level

CT : cervicothoracic, T : thoracic, LS : lumbosacral, LMMC : lipomyelomeningocele, MMC : myelomeningocele, SCM : split cord malformation

admission.

3) We checked the radiographic findings of CDS in plain X-rays, ultrasonography, CT and MRI's.

4) We checked the patients' pathological profiles including the terminal point, combined anomalies and combined tumors. Through this, we want to verify the statement that "Sacrococcygeal dimples have no connections with neural structures so there is no need to evaluate"<sup>15)</sup>.

5) Complete removal is known as the best treatment strategy. But in cases whom incomplete removal was inevitable, we had to remove the sinus tract in the range until squamous epithelium was not exist. We checked whether that could be a supplementary strategy.

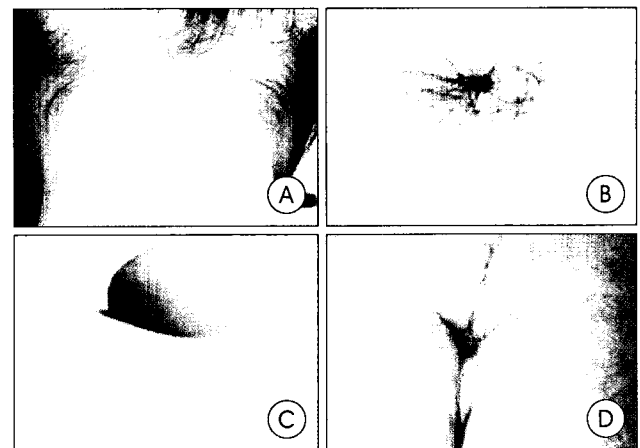
6) We checked the patients' postoperative courses, including newly developed neurological deficits.

## Results

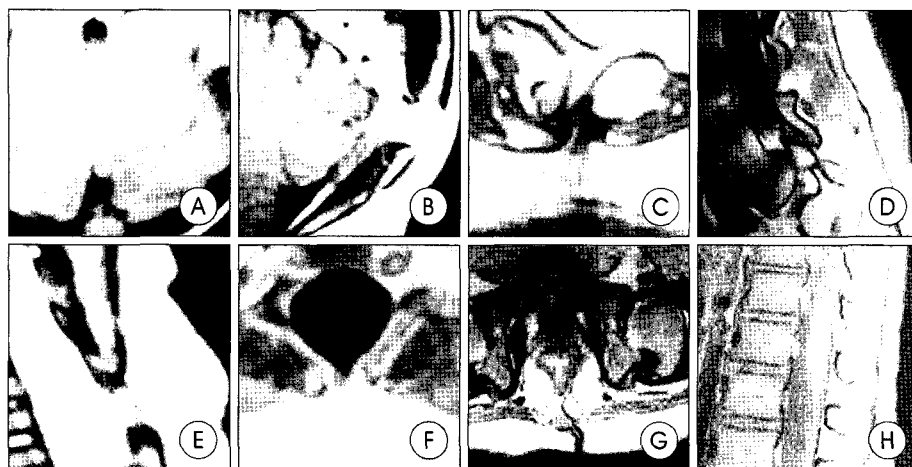
### Timing of diagnosis

Because CDS is a congenital lesion, a cutaneous pit must be diagnosed at birth. But it's not easy to find such a tiny little pit on the newborn baby. However, in cases that : 1) the cutaneous pits except on the sacrococcygeal area, 2) the cutaneous pits on sacrococcygeal area along with other skin lesions (ex.

hypertrichosis, cutaneous hemangioma, cysts, and subcutaneous lipomas), and 3) a patient showing discharge on the pit or meningeal irritation signs; one must perform the evaluations to diagnose CDS. Strikingly only 6 out of 20 cases were diagnosed and treated in time. Four of 6 showed no neurological deficits, 1 showed an improvement in weakness and 1



**Fig. 1.** A shows a midline cutaneous pit situated on the suboccipital area. B shows a hypertrichosis and hemangioma around the cutaneous pit. C shows cystic mass(meningocele). In this case we diagnosed congenital dermal sinus postoperatively because the cutaneous pit was hidden around the neck of the cyst. D shows subcutaneous lipoma and pit.



**Fig. 2.** A & B show a dermoid cyst and skull defect at the suboccipital area. C, D, E, F, G & H show a low signal intensity subcutaneous tract which is continuous to the intraspinal space but its end point is not well delineated.

required urinary catheterization postoperatively, but we thought it as a result of combined lipoma.

Thirteen out of 20 cases were not diagnosed within a preferred time period. From meaningful symptoms to diagnosis took an average of 2 years 4 months. The reasons for delayed diagnosis included both the fault of clinicians (patient observation, missed CDS symptoms when operating on combined anomalies and not considering CDS in recurrent meningitis ect.) and that of families (arbitrary observation, and refusing the clinician's recommendation to implement ect.). Seven of 13 showed no neurological deficits, 4 showed improvements in weakness and 2 showed voiding problems which may have been due to lipoma. 1 case combined with lipoma had its opening not in the midline and was excluded. CDS has only been located in the midline so far.

### Clinical manifestations

Fourteen patients had skin manifestation as their chief complaint. One complained of meningeal irritation symptoms. Four cases referred to paralysis. One case has been discovered incidentally in the course of working with another disease. Skin manifestations in the cutaneous pit included: mass (8 cases), discharge (11 cases), hypertrichosis (6 cases), hemangioma (3 cases), dysplastic skin (1 cases). The natures of the mass are 4 subcutaneous lipomas, 2 meningoceles, 1 subcutaneous dermoid tumor and 1 un-specified mass presumed to be an abscess(Fig. 1). The discharge cases included pus (7), keratin material (2) and 2 of an unspecified nature. Eleven cases showed no neurological deficits, 6 cases showed voiding or defecation difficulties, 4 cases showed various weaknesses, and 5 cases showed orthopedic abnor-

malities including scoliosis.

In the group that did not have lipoma combinations (15 cases), 9 cases had no neurological deficits, 3 cases had voiding or defecation difficulties, 4 cases had weaknesses, and 3 cases had orthopedic abnormalities.

Three cases showed spinal abscesses at admission. Five cases showed meningitis, and 1 case had a previous meningitis history.

### Radiologic findings

Seven cases had problems located in the occipital area. The radiographics of 5 patients showed that they had bony defects in the occipital area. The other 2 had posterior arch defects on the atlas. Thirteen cases were located in the spinal area. We obtained the radiographs of 10 of these patients, and we found that 4 of them had no radiologic abnormalities. Two cases combined with Currarino's triad had sacral agenesis, while 2 spinal bifida and 2 hemivertebrae were detected on plain X-rays. A preoperative ultrasonogram had been performed on two of the orbital cases. One case, was combined with encephalocele, and had meningocele. We couldn't confirm the connection between the sinus tract and neural structures. Four spinal cases had a preoperative ultrasonogram performed as well, and we were able to obtain information about combined anomalies, combined tumors, level of conus medullaris and cord pulsation, but we couldn't confirm the connection between the sinus tract and neural structures. Two occipital cases had CT scans performed, but not MRI's. MRI's had not yet become popular at that time, so we could only confirm the combined tumors and hydrocephalus through CT scans. Except for the 2 cases mentioned above, 18 cases had MRI's performed. An MRI is considered as a 'gold standard' diagnostic tool for CDS. We can confirm combined tumors, combined anomalies, level of conus and we can even trace the sinus tract through a MRI. Within the spinal canal, however, we can't trace the sinus tract yet(Fig. 2).

### Pathologic profiles

CDS locations were classified by its' internal placements. Nine cases were located in the lumbosacral area, 7 cases were in the occipital area, 2 cases were in the thoracolumbar area, 1 case was in the cervicothoracic area, and 1 case was not

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clearly classified because of a wide spread infection. Four lipomyelomeningocele(LMMC), 2 Currarino's triad, 1 encephalocele, 1 myelomeningocele and 1 diastematomyelia were combined with CDS. Eleven dermoid tumors and 1 suspicious ruptured dermoid tumors were combined with CDS. One dermoid tumor suspected from a radiogram was not confirmed pathologically. Four lumbosacral located CDS openings were low as noted in the sacrococcygeal dimple discussed previously.

### Surgery

Total removal was possible in 16 cases. In 2 cases, the combined dermoid tumors were adhered to neural structures so we could only partially remove it and the remaining capsule was coagulated. Because of a wide spread infection, we couldn't find the internal ending of the sinus in one case, so we could only partially remove the tract. We were able to confirm that no pathological squamous epithelium remained in the stump. No recurring problems in that case have resurfaced. In one case, the sinus tract was mingled with the venous sinus. We did a subtotal resection and confirmed that no squamous epithelium remained. That case has also had no new problems.

### Postoperative course

None of the cases developed new neurological deficits postoperatively. Seven cases showed urination or defecation problems preoperatively. They were all combined with LMMC or nervous system infections. In cases where CDS was combined with LMMC, the clinical course was similar to that of LMMC. Four paralysis cases showed improvements in weakness postoperatively. Two of them are capable of living independent lives.

## Discussion

**CDS** is a not so rare congenital disease that results from the failure of the neuroectoderm to separate from the surface ectoderm during the process of neurulation<sup>5,11</sup>. It can also develop from the accessory neurenteric canals that remain<sup>10,12</sup>. It is known that it can develop only in the midline of the body. CDS has a midline cutaneous pit that is covered with squamous epithelium.

It connects variously with the spinal cord, spinal nerve, arachnoid, and the dura or muscles<sup>8</sup>. If there is a connection between the sinus and neural structures, a skin infection can spread throughout the nervous system. A physical examination only will barely detect the internal ending, and the use

of probes or fistulagrams are not allowed<sup>6</sup>. Because CDS has the ability to spread existing infections, MRI can be a golden diagnostic tool. A MRI can visualize the sinus tract, but its validity is limited in the intraspinal space<sup>7,13</sup>. In surgical timing, the prognosis of patients' that were treated in time was not superior to that of patients' that were treated at a later date. It does not mean, however, that the statement "a CDS patient must be operated as soon as it diagnosed." is not true. It simply means that CDS prognosis is largely dependant on the nervous system infection. The prognosis of a patient who has a nervous system infection is worse than that of one with a recurrent cutaneous infection.

It's not easy to predict whether a cutaneous infection will progress to a nervous system infection or not. Considering the poor outcome of nervous system infections, it makes sense that CDS should be treated at diagnosis<sup>5,14</sup>. All cases showed cutaneous abnormalities at admission and skin manifestations are the most common chief complaint<sup>1,3,4</sup>. Theoretically all CDS patient showed cutaneous pit from birth. But in our series we couldn't find cutaneous pit preoperatively in 4 cases. In 3 cases sinus openings were hide beneath the neck of mass and in 1 case opening was unclear because of previous operation. As long as the sinus tract was located lower than the level of 3rd sacrum or intergluteal fold, or directed downward toward the caudal side, it is considered as a sacrococcygeal dimple. Physicians thought that they would need no further diagnostic evaluations except physical examinations<sup>15</sup>.

If, however, they were combined with other skin manifestations that presented occult spinal dysraphisms, they would need further radiologic evaluations. The sinus tract of CDS can also communicate with neural structures when they are located below the normal level (L1-2 level). Combined anomalies from our studies have already been mentioned in other articles. Combined tumors were all dermoid tumors in our observations, and are consistent with previous studies<sup>5</sup>. If there is a nervous system infection, it can be disastrous. In this case, CDS must be surgically removed immediately once it has been diagnosed<sup>5,16</sup>. But if it is mingled with the spinal cord or venous sinus, total removal is impossible. In that case we remove as much of the sinus tract as possible and do a frozen biopsy on the remaining stump. If no squamous epithelium remained at the stump, we ended the surgery. In these cases, no recurrence of pathology was detected as happens in total resections. Combined dermoid or epidermoid tumors must be surgically removed totally, because remaining tumors can be a source of infection or tumors can from it<sup>9</sup>.

## Conclusion

We can verify the validity of the statement that "CDS must be surgically removed immediately if they are diagnosed". MRI has the key role in diagnosing CDS, but it can't visualize the sinus tract in the intraspinal portion yet.

"The Sacrococcygeal dimple that is located in the sacrococcygeal area almost invariably have no connection with intraspinal structures", should be revised as "Sacrococcygeal dimple almost invariably have no connection with intraspinal structures if they have no other cutaneous manifestations suggesting occult spinal dysraphism.

"Total Surgical removal is the principle treatment of CDS" is still valid. In cases where total removal is impossible, the "Resection in the area where no squamous epithelium remains" can be an alternative treatment, but it needs further investigation.

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