Surgical implications of the thickened pituitary stalk accompanied by central diabetes insipidus

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Object. The authors discuss the indications for and timing of a diagnostic neurosurgical procedure in children with diabetes insipidus (DI) and a thickened pituitary stalk (TPS) on magnetic resonance (MR) imaging.

Methods. Seven children with a TPS who presented with DI eventually underwent surgery for diagnostic purposes. The ages at onset of DI were 6 to 16 years, and the follow-up period until surgery was 26.9 ± 11.9 months. In four of seven children, the stalk appeared normal on the first MR image, but it was thickened and variably enhancing on later images in all instances. The reason for eventual surgery was endocrinological deterioration in two of seven children, radiological progression in two children, and a combination of the two in three children. Three children experienced visual disturbances and four children had optic nerve, chiasma, or hypothalamus involvement. All children suffered additional endocrinological abnormalities pursuant to the initial DI. A definitive diagnosis was achieved in six of seven children: germinomas in five and Langerhans cell histiocytosis in one. One child had lymphocytic infiltrate. None of the children deteriorated neurologically or endocrinologically after the operation. On follow up, vision deficit was irreversible in all children who demonstrated visual abnormalities before treatment.

Conclusions. Surgery should be performed in children with a TPS and DI for early diagnosis and disease-oriented therapy when there is further endocrinological, radiological, or clinical deterioration. The complication rate is low in open biopsies, and histological diagnosis is achieved in most of the cases. All children who present with central DI must undergo head MR imaging, and even if results are normal, close radiological and clinical follow up is mandatory.

Key Words • pituitary stalk • brain tumor • biopsy • diabetes insipidus • magnetic resonance imaging • pediatric neurosurgery

Diagnostic surgery in a child presenting with DI and a TPS on MR imaging is a confusing issue for the treating pediatrician, endocrinologist, radiologist, and neurosurgeon. This situation may represent a wide differential diagnosis, including neoplastic, inflammatory, autoimmune, infectious, or malformative processes. The natural history of the disease in a particular child may therefore not be anticipated on the basis of radiological appearance or endocrinological deficiency. Isolated TPS by itself can also represent a wide differential diagnosis, including rare cases of tuberculosis, sarcoidosis, and the more common pathological entities of LCH, lymphocytosis, and germ cell tumors. Anecdotal cases of a previously diagnosed histiocytosis transforming to a germinoma have also been reported.

When the diagnosis is not evident, the issue of biopsy is usually raised, but in most of the cases it is rejected. The patients are then given substitution hormonal therapy and are followed up by repeated MR imaging or CT scanning. During the follow-up period, however, some children will deteriorate clinically, manifesting growth arrest because of GH deficiency and will require additional hormonal therapy or increasing doses of steroid agents. Such deterioration can be attributable to a tumoral disease, such as germ-cell spread, even when tumoral markers are negative. In some cases, diagnosis and disease-oriented therapy will be delayed many months after the onset of DI and appearance of TPS.

In the clinically stable child, surgery might seem too aggressive, yet the following questions must be raised when dealing with this subject: 1) Will surgery change the treatment of the child with DI and TPS? 2) In the absence of radiological progression, is a child with significant or progressive endocrinological deficit still considered a “stable” patient? 3) What would be the yield of a definitive diagnosis in case a biopsy is performed? 4) What is the risk–benefit ratio of diagnostic surgery?

Authors of some series have discussed the differential
diagnosis, radiological changes, and endocrinological features of children with DI and TPS, but the indications to perform a diagnostic surgery, its timing, and the related risk–benefit balance are not clearly addressed in the literature. In this paper, we report on seven children presenting with DI and TPS who underwent surgery for diagnostic purposes. We then discuss the surgical implications of this pathological entity, analyzing our data together with information provided by previously reported series.

Clinical Material and Methods

Patient Population

The present series describes seven children with central DI who also presented with a TPS on MR imaging (Figs. 1 left, 2a, and 3) and underwent surgery at Necker Children’s Hospital (Paris) or Dana Children’s Hospital (Tel-Aviv) between 1993 and 2000. The arguments that led to the diagnostic surgeries are set forth, together with the clinical, endocrinological, and radiological data (Tables 1–3). Postoperative treatment and follow up are also detailed (Table 1). All patients had good-quality MR images, which included T₁-weighted images with and without contrast, as well as sagittal, coronal, and axial cuts. Parameters for CSF were evaluated in all patients to exclude infectious causes. Endocrinology studies included measurements of stimulated GH levels, cortisol levels (morning basal level and during insulin-induced hypoglycemia), free T4 and thyroid-stimulating hormone levels, and gonadotropin levels. All children were evaluated by an endocrinologist.

Results

Pathological and Laboratory Findings

Of the seven children, five were diagnosed as suffering from germ cell tumors (57.1%), one suffered from histiocytosis (14.3%), and one had a nonconclusive diagnosis in which the histological studies showed lymphocytic infiltrate, with polynuclear eosinophils (Table 1). In this patient, several large cells were observed, without mitoses or atypia, so that the cytological aspect was not compatible with a germinoma. He is being followed up and is receiving hormone therapy.

Tumor markers, including β–human chorionic gonadotropin and α-fetoprotein, and cytology for CSF were studied systematically; however, these were negative preoperatively in all patients. Cultures of CSF, cell count, protein, and glucose were obtained in all children. All cultures were negative, and none of the patients’ CSF parameters were suggestive of infection, including TB. The child with the nonconclusive diagnosis (Case 1) had a negative TB test.

Clinical and Endocrinological Features

All children (five girls and two boys) presented with central DI. The mean age at diagnosis was 10.4 ± 3.2 years (range 6–16 years). The mean follow-up period between diagnosis of DI until surgery was 23 ± 15 months (Table 2), and the age at surgery was 12 ± 3.8 years (range 9.4–20 years).

During the preoperative follow-up period, all children were treated with desmopressin acetate for DI (some needed increasing doses); by the time they underwent surgery, they were receiving additional hormone therapy, including hydrocortisone (five patients), L-thyroxine (two patients),
and ethynil-estradiol (one patient). Four children presented with slowed or arrested growth, mainly affecting the stature curve. A GH deficiency was noted in five children. Three of the seven children had significant persisting headache.

None of the children had visual abnormalities on presentation with DI, but three experienced quadranopia (one patient) or bitemporal hemianopia (two patients), two of them demonstrating a rapidly progressing visual loss, with functional blindness in Case 6. These visual deficits were irreversible, despite optimal disease-oriented treatment after surgery and obvious remission of the disease on MR imaging.

Radiological Features

The earliest radiological abnormalities, as detected on MR imaging, were TPS and absence of a hyperintense signal of the posterior hypophysis in three children. On subsequent MR imaging studies, progression of the disease caused infiltration of optic nerves, chiasma, or hypothalamus in four patients (Table 3). Preoperative radiological deterioration appeared in five children: four were diagnosed as having germinomas and one as having LCH. Radiological remission of disease was achieved in all patients who were treated for germinoma and in the patient with LCH (who received both radiotherapy and chemotherapy).

Surgical Aspects and Outcome

The reasons for surgery for each child are presented in Table 1: radiological deterioration in five children and clinical deterioration (increasing use of hormone therapy because of progressive deficits or visual disturbance) in four children. The patient in Case 1 suffered a significant growth arrest for more than 2 years (−3 standard deviations) and was a candidate for GH therapy. Before initiation of such treatment, tissue diagnosis to rule out tumoral disease was decided on. The patient in Case 7 was sent from another country in request for reevaluation for radiotherapy treatment for what was considered at another institution as a hypothalamic glioma presenting as TPS.

All children underwent craniotomies (six pterional, one subfrontal). No intra- or postoperative complications occurred. From an endocrinological point of view, there was

![FIG. 3. Case 1. Coronal and sagittal MR images of TPS in a child with DI.](image)

<table>
<thead>
<tr>
<th>TABLE 1</th>
<th>Initiative for operation, symptoms, and pathology of children with DI and TPS*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case No.</td>
<td>Reason for Op</td>
</tr>
<tr>
<td>1, M</td>
<td>large PS, endocrine deficit, enable GH treatment?</td>
</tr>
<tr>
<td>2, F</td>
<td>clinical &amp; radiological deterioration</td>
</tr>
<tr>
<td>3, M</td>
<td>PS changes &amp; invasive process into 3rd ventricle</td>
</tr>
<tr>
<td>4, F</td>
<td>clinical &amp; radiological deterioration</td>
</tr>
<tr>
<td>5, F</td>
<td>clinical &amp; radiological deterioration</td>
</tr>
<tr>
<td>6, F</td>
<td>clinical &amp; radiological deterioration</td>
</tr>
</tbody>
</table>
Surgical implications of the thickened pituitary stalk

Clinical and endocrinological features of children undergoing surgery for TPS and DI*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Time of FU</th>
<th>Markers (CSF &amp; Blood)</th>
<th>GH</th>
<th>LH</th>
<th>FSH</th>
<th>TSH</th>
<th>PRL</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>At DI</td>
<td>At Op</td>
<td>After DI (mos)</td>
<td>dec</td>
<td>dec</td>
<td>norm</td>
<td>norm</td>
<td>norm</td>
</tr>
<tr>
<td>1</td>
<td>8.5</td>
<td>9.4</td>
<td>11</td>
<td>neg</td>
<td>dec</td>
<td>norm</td>
<td>norm</td>
<td>norm</td>
</tr>
<tr>
<td>2</td>
<td>12</td>
<td>14.1</td>
<td>24</td>
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<td>dec</td>
<td>norm</td>
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</tr>
<tr>
<td>3</td>
<td>8.5</td>
<td>10.8</td>
<td>4</td>
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<td>norm</td>
</tr>
<tr>
<td>4</td>
<td>10</td>
<td>11.5</td>
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<td>dec</td>
</tr>
<tr>
<td>5</td>
<td>16</td>
<td>20.0</td>
<td>48</td>
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<td>norm</td>
</tr>
<tr>
<td>6</td>
<td>12.0</td>
<td>14.0</td>
<td>24</td>
<td>neg</td>
<td>dec</td>
<td>norm</td>
<td>norm</td>
<td>norm</td>
</tr>
<tr>
<td>7</td>
<td>6.0</td>
<td>9.0</td>
<td>36</td>
<td>neg</td>
<td>norm</td>
<td>norm</td>
<td>dec</td>
<td>norm</td>
</tr>
</tbody>
</table>

* Abbreviations: CORT = cortisol; dec = decreased; FSH = follicle-stimulating hormone; FU = follow up; inc = increased; neg = no aggravation after surgery, and children remained on hormone therapy consistent with their endocrinological profiles. Adjuvant therapy was given in six children, with remission of the disease in five (the patient who received a diagnosis of LCH received vinblastine, prednisone, and limited radiotherapy at a dose of 12 Gy; four children were treated with the accepted protocols for germ tumors, and one is currently under treatment). Postoperative follow up for the whole group ranged from 2 months to 7 years (Table 1). None of the children who had preoperative visual problems experienced improvement in their vision despite successful disease control. The patient in Case 6 was practically blind at time of surgery and remained so after treatment, with full remission of her oncological disease.

Discussion

A TPS in a child presenting with DI may be the first radiological sign of a progressive disease, such as a germ-cell tumor or LCH. These diseases can cause irreversible damage while the children are being treated conservatively in the absence of an accurate diagnosis. Three of the children in the present series experienced progressive visual disturbance that was irreversible even after treatment, and all children had additional hormone deficits pursuant to the initial DI.

The seven children reported on in this paper were categorized, in the first stages of their disease, as idiopathic cases of central DI, because no evident disease was discerned in blood, CSF, and other tests, and because initial brain MR images or CT scans were interpreted as normal. Obviously, the term “idiopathic DI” should be the diagnosis of exclusion, and must be reviewed during close follow up, because certain diseases may manifest months or even years after the onset of DI. The differential diagnosis of a central DI accompanied with or followed later by a TPS is wide, including infectious (TB), inflammatory (sarcoidosis), and tumoral processes. Nevertheless, in up to 25% of children, no organic cause can be identified. In places where TB is a major concern, thorough investigation should include chest x-ray studies, tuberculin tests, and even polymerase chain reaction analysis before a neurosurgical procedure is undertaken. In non endemic places, however, CSF parameters as well as final culture (with specific emphasis on TB if parameters such as glucose or cell count are abnormal) may suffice, unless there is high suspicion for a particular patient.

In most centers, a child with TPS and DI will be treated initially by hormonal therapy only. A tissue biopsy is usually delayed until a progressive lesion is recognized on subsequent MR imaging studies. On the data presented in this paper, however, as well as in previous publications, we emphasize the importance of an early diagnostic surgery. With tissue diagnosis, an early specific treatment is possible. This approach may avoid loss of vision, which can be rapid, progressive, and sometimes irreversible. Also, further endocrinological deterioration—including development of panhypopituitarism—can be prevented. In cases in which GH therapy may be beneficial, it might be important to verify whether the underlying disease is tumoral or not, especially for physicians who believe that GH therapy must not be given in the presence of an untreated tumor.

Early diagnosis in the case of germinoma may enable treatment before metastatic disease occurs, thus avoiding an aggressive protocol that includes total-spine irradiation. The danger of a sudden metastatic disease in a child followed up for DI and TPS is not theoretic. A 7-year-old girl presented initially with DI to another hospital (unpublished case). The first MR images demonstrated TPS (Fig. 2a). Three months later, abnormal contrast uptake was noted in the stalk area but no other lesions were detected, and the child had no additional symptoms or signs. Magnetic resonance imaging performed 6 months later, however, showed a clear progressive disease, with metastatic spread into the spinal canal (Fig. 2b–d). At this point, the child manifested mild ataxia, as well as some visual problems that occurred abruptly. Repeated tests showed that levels of β-human chorionic gonadotropin were 42 ng/ml and 0.19 ng/ml in the CSF and blood, respectively, and that α-fetoprotein remained normal. A lumbar puncture disclosed germinoma cells.

Even though it is hard to estimate how many children with TPS and DI will have germ-cell tumors, one can conclude that this situation is not rare. In our series, five (71.4%) of the seven children had germ-cell tumors. Mootha et al. presented a series of nine children with central DI, seven of whom demonstrated TPS. Biopsies were obtained, and six tumors were diagnosed as germinomas. The delay between diagnosis of DI or TPS and the surgical procedure, if performed, is usually months to years. The main reasons for the delay has been the belief that surgery was not needed in most of the cases, and if performed, might have unnecessary complications. There is also the

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TABLE 2

"No aggravation after surgery, and children remained on hormone therapy consistent with their endocrinological profiles. Adjuvant therapy was given in six children, with remission of the disease in five (the patient who received a diagnosis of LCH received vinblastine, prednisone, and limited radiotherapy at a dose of 12 Gy; four children were treated with the accepted protocols for germ tumors, and one is currently under treatment). Postoperative follow up for the whole group ranged from 2 months to 7 years (Table 1). None of the children who had preoperative visual problems experienced improvement in their vision despite successful disease control. The patient in Case 6 was practically blind at time of surgery and remained so after treatment, with full remission of her oncological disease.

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theoretical possibility that the underlying disease might be self-limited, given the fact that the differential diagnosis is wide, and the natural history is thus unpredictable in the particular child.

Interestingly, five of the seven patients were girls, and most of them had germ cell tumors. The authors suggest that TPS may be an underdiagnosed pathological entity, especially in girls, because precocious puberty could be ignored in girls more than it is in boys. Also, not all children undergo MR imaging on presentation of endocrinological problems; in addition, the fine changes in the thick stalk may be overlooked by a CT scan.

Endocrinological Considerations

Central DI is usually a chronic condition, in which vasopressin levels are typically low. It may present months or even years before other endocrinological or radiological abnormalities develop. By the time children arrive to surgery, however, additional endocrinological abnormalities are observed. These may include GH deficiency, with significant slowing or arrest of growth (mainly stature), gonadotropin and thyrotropin deficiency, and hyperprolactinemia. In our patients, four presented significant growth attenuation, with very low or undetectable GH levels. This situation differs from the one of idiopathic GH deficiency described previously by Pinto, et al., in which among 15 children of pubertal age with GH deficiency, 60% had gonadotropin deficiency. None of these children had DI, and in most of the cases the diagnosis of GH deficiency was made before the age of 2 years.

Leger, et al., described 26 children with central DI and TPS on MR imaging. Endocrinological analysis of these children led to classification in three different groups: isolated central DI, DI and GH deficiency, and GH deficiency with at least one other anterior pituitary hormone abnormality. All children with isolated DI were idiopathic; however, the second and the third groups included germinomas, histiocytosis, and idiopathic cases. The highest incidence of germinomas was found in the third group. Multiple hormonal deficiency pursuant to DI may indicate a progressive disease and is commonly found in patients with a suprasellar mass invading the PS. It is clear, however, that one cannot make a definite diagnosis of the underlying disease by the endocrinological profile alone, because multiple-hormone deficiency has been described in germinomas, as well as in LCH and lymphocytic hypophysitis.

Radiological Aspects

Currently, it accepted that MR imaging is the preferred radiological investigation in a patient with central DI. It is not clear how many children with central DI will have abnormal MR images, but the estimation is that when the MR image is carefully read, with the specific question referred to the PS and posterior hypophysis, the yield is approximately 60 to 70% on initial studies, and higher on subsequent ones. At the early stages of LCH or germinoma, the MR image may seem normal. Specific attention should be paid, however, to absence of hyperintensity of the posterior hypophysis on unenhanced T1-weighted images, and to the appearance of a TPS. These features may be the first or the only radiological signs of the underlying disease.

Mootha, et al., reported on a series of nine consecutive children with central DI who underwent brain MR imaging on presentation. Five of nine studies were originally reported as abnormal, whereas on revision as many as seven (77.8%) were read as abnormal. By 14 months of follow up, however, all MR imaging studies showed positive findings. The first abnormal finding in all patients was isolated PS thickening. If the disease is self-limited, it is likely that subsequent images would show regression of the thickening. In progressive cases, the disease can spread to adjacent areas such as the hypothalamus, optic nerves, and chiasma, invade the ventricular system, or cause radiologically evident metastatic disease. Thus, when definitive tissue diagnosis is not available, repeated MR imaging studies are mandatory.

Neurosurgical Aspects

The timing and indication of surgery in a child with DI and TPS are still unsettled issues. One may assume that if a diagnosis is achieved earlier and proper therapy is then given, clinical deterioration can be minimized. This argument may be correct not only for germinomas, where the tumor-oriented therapy cannot be initialized until a definitive diagnosis is made, but also in resistant cases of LCH. In some cases of LCH and hypophysitis, chemotherapy or radiotherapy are indicated, as was the case in one patient in the present series and was reported in previous papers. In rare cases, several biopsies along the time of follow up may be needed, because hypophysitis can progress or transform into a tumor. Bettendorf, et al., reported a case of a young girl whose central DI and panhypopituitarism evolved over a period of 10 years. Two years after the DI was diagnosed, MR imaging demonstrated a TPS, which progressed to pituitary enlargement and compression of the optic chiasma. A transsphenoidal biopsy procedure was performed, and the histological specimen showed active hypophysitis with lymphocytic infiltrates and necrosis. High-dose dexamethasone was followed by stereotactic radiation therapy, with significant improvement on MR imaging. Subsequent MR imaging showed significant improvement on MR imaging. Subsequent MR imaging showed significant improvement on MR imaging.

### TABLE 3

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Initial MR Study</th>
<th>Preoperative MR Study</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>absence of hyperintense signal of posthypophysis†</td>
<td>TPS</td>
</tr>
<tr>
<td>2</td>
<td>absence of hyperintense signal of posthypophysis &amp; TPS</td>
<td>inc TPS</td>
</tr>
<tr>
<td>3</td>
<td>thick &amp; enhancing PS</td>
<td>TPS &amp; lesion invading the infundibulum of 3rd ventricle</td>
</tr>
<tr>
<td>4</td>
<td>TPS†</td>
<td>TPS &amp; enhancing mass extending to optic chiasm</td>
</tr>
<tr>
<td>5</td>
<td>absence of hyperintense signal of posthypophysis†</td>
<td>TPS &amp; enhancing lesion extending into optic nerve</td>
</tr>
<tr>
<td>6</td>
<td>norm†</td>
<td>TPS &amp; chiasmatic thickening</td>
</tr>
<tr>
<td>7</td>
<td>TPS</td>
<td>TPS</td>
</tr>
</tbody>
</table>

* Patients in Cases 1 and 7 had only one preoperative MR imaging study (but underwent a previous CT scan).
† Normal CT scan.

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imaging disclosed several intracranial lesions, however, which were later identified histologically as a germinoma.

The other surgical consideration is the operative technique used to obtain biopsy specimens. The surgical approach, whether open biopsy through craniotomy or transsphenoidally, depends on several factors: the preference of the surgeon, the size and location of the lesion, the presence of adjacent or other intracranial lesions, and the age of the patient.

We believe that the choice of open biopsy through a craniotomy is the preferred method in children with TPS. In the cases detailed in this report, ptetional and subfrontal-transglabellar approaches were used. These methods enable us to appreciate the macroscopic look of the pathological entity (that is, its color and consistency). The biopsy specimen can be safely taken even from very small lesions, and the optic nerves, chiasm, and PS are clearly visualized. Sometimes, intraoperative image guidance is helpful. In the presence of larger lesions, especially when the tissue is somewhat necrotic, several biopsies can be performed if necessary. These approaches are feasible in very small children as well as in the older ones. The complication rate is low, and in experienced hands no new neurological deficit should usually be expected. Other authors may prefer the transsphenoidal biopsy. Its advantage lies in the avoidance of an intracranial operation, but it may be technically difficult in very small children, for very small lesions, or for an isolated TPS (in which case, it might be difficult to detect the proper site for a biopsy).

Conclusions

As long as a definite diagnosis is not available, it is mandatory that the child with DI is followed closely both clinically and radiologically, and that MR imaging should be repeated systematically, even if the initial studies were interpreted as normal. This procedure is most relevant when additional endocrinological deficiencies are observed.

The low rate of surgical complications, the high rate of definitive diagnosis, and the chance of optimizing treatment all favor early diagnostic surgery in the child with TPS and DI. The appropriate timing would be when either clinical, radiological, or endocrinological progression is evident. In the case of endocrinological or clinical aggravation, we do not believe that one should necessarily wait until further radiological progression appears on MR imaging. Indeed, in the era of modern neurosurgery, the risk of surgery may be less than the risk of irreversible visual or endocrinological deficit.

References


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