

# The Presentation and Management of Nasal Dermoid

## A 30-Year Experience

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**Objective:** To review the presentation of nasal dermoid in children and present guidelines for its management.

**Design:** Retrospective study (January 1, 1970, through December 31, 2000).

**Setting:** Tertiary-care pediatric medical center.

**Patients:** Number of patients: 42 (28 boys and 14 girls).

**Intervention:** Extensive review of the initial presentation, significant family and medical history, workup, surgical approach, complication, and rate of recurrence.

**Results:** Mean age of presentation was 32 months. The most common presentation was a nasoglabellar mass, in 13 patients (31%). Five patients presented with an as-

sociated craniofacial abnormality. Thirty-nine patients (93%) underwent a preoperative imaging workup. Thirty-one (74%) did not show any clinical and/or radiographic indication of intracranial extension. Thirty-four (81%) underwent extracranial excision, and 8 (19%) underwent combined intracranial-extracranial excision. Five patients (12%) presented with recurrence, extracranially in 4 and intracranially in 1. No other complication was noted, with a mean follow-up of 7 years.

**Conclusions:** Nasal dermoid is a rare congenital anomaly. Preoperative evaluation is essential to rule out intracranial extension. Surgical strategy depends on the location and extent of the lesion, ranging from local excision to a combined intracranial-extracranial approach. Recurrence is uncommon and often easily managed.

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**N**ASAL DERMOID is a rare developmental anomaly. Unlike other craniofacial dermoids, the nasal lesions can present as a cyst, a sinus, or a fistula and may have an intracranial extension.<sup>1</sup> The incidence is estimated at 1:20 000 to 1:40 000 births.<sup>2,3</sup> Pathogenesis involves the incomplete obliteration of neuroectoderm in the developing frontonasal region.<sup>2,4</sup> Progressive enlargement of a nasal dermoid can cause soft tissue and skeletal deformity, local infection, meningitis, and brain abscess. Timely diagnosis is essential, and surgical excision is the only therapeutic modality.

The purpose of this study is to review a 30-year experience in the management of nasal dermoids at The Children's Hospital, Boston with emphasis on the presentation, role of preoperative imaging, surgical approach, and rate of recurrence. On the basis of our findings, we present a treatment algorithm for the management of nasal dermoid.

## METHODS

One hundred fifty patients with craniofacial dermoids were treated at The Children's Hospital, Boston from January 1, 1970, through December 31, 2000. Only patients with a nasal dermoid, complete medical record, and sufficient follow-up time were included in this study. We recorded data including the child's initial presentation, significant medical and family history, preoperative imaging findings, surgical procedures, intraoperative findings, and complications, including recurrence. We included 42 patients (**Table**) in this study (28 boys [67%] and 14 girls [33%]). The age at presentation ranged from 1 month to 16 years (mean age, 32 months).

## RESULTS

The most common presentation was a nasoglabellar mass (n = 13 [31%]). Other presentations included a dorsal mass (n = 11 [26%]), dorsal sinus (n = 8 [19%]), tip fullness (n = 4 [10%]), dorsal sinus with hair (n = 2 [5%]), dorsal widening (n = 2 [5%]), bifid tip (n = 1 [2%]), and lateral orbital

**Patient Characteristics and Management of Nasal Dermoid\***

| Patient No./ Sex/Age, mo | Associated Problems          | Initial Presentation | Imaging     | POIE | Initial Procedure                  | IOIE | Recurrence, Postoperative Years | Location of Recurrence | Subsequent Procedure | Follow-up, y |
|--------------------------|------------------------------|----------------------|-------------|------|------------------------------------|------|---------------------------------|------------------------|----------------------|--------------|
| 1/F/8                    | None                         | Dorsum mass          | Facial film | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 10           |
| 2/M/17                   | None                         | Glabella mass        | CT          | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 11           |
| 3/M/30                   | None                         | Nasal dorsum mass    | CT          | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 13           |
| 4/M/10                   | None                         | Glabella mass        | CT and MRI  | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 10           |
| 5/M/48                   | Hemifacial microsomia        | Dorsum sinus         | Facial film | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 10           |
| 6/M/2                    | None                         | Dorsum mass          | None        | ...  | Vertical excision                  | No   | ...                             | ...                    | ...                  | 10           |
| 7/M/2                    | None                         | Dorsum mass          | Facial film | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 15           |
| 8/F/48                   | None                         | Dorsum mass          | Facial film | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 10           |
| 9/M/8                    | None                         | Dorsum mass          | MRI         | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 8            |
| 10/M/6                   | None                         | Dorsum sinus         | MRI         | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 8            |
| 11/M/8                   | None                         | Glabella mass        | CT          | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 5            |
| 12/M/66                  | None                         | Widened dorsum       | CT and MRI  | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 6            |
| 13/M/12                  | None                         | Dorsum mass          | None        | ...  | Vertical excision                  | No   | ...                             | ...                    | ...                  | 7            |
| 14/F/24                  | None                         | Glabella mass        | CT          | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 2            |
| 15/M/24                  | None                         | Glabella mass        | CT          | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 5            |
| 16/F/1                   | Bilateral lacrimal duct cyst | Nasal tip fullness   | CT and MRI  | No   | Vertical excision                  | No   | 2                               | Intracranial           | Frontal craniotomy   | 2            |
| 17/F/24                  | None                         | Dorsum sinus         | CT          | No   | Vertical excision                  | No   | ...                             | ...                    | ...                  | 6            |
| 18/M/18                  | None                         | Dorsum sinus, hair   | CT          | Yes  | Vertical excision                  | No   | ...                             | ...                    | ...                  | 5            |
| 19/M/26                  | None                         | Dorsum sinus         | CT          | No   | Transverse excision                | No   | ...                             | ...                    | ...                  | 8            |
| 20/F/29                  | None                         | Glabella mass        | CT          | No   | Transverse excision                | No   | ...                             | ...                    | ...                  | 7            |
| 21/F/192                 | None                         | Dorsum sinus/hair    | CT          | No   | Transverse excision                | No   | ...                             | ...                    | ...                  | 10           |
| 22/M/12                  | None                         | Dorsum sinus         | CT          | No   | Transverse excision                | No   | ...                             | ...                    | ...                  | 5            |
| 23/F/24                  | None                         | Glabella mass        | CT          | Yes  | Transverse excision                | No   | ...                             | ...                    | ...                  | 8            |
| 24/M/6                   | None                         | Dorsum sinus         | MRI         | No   | Lateral rhinotomy                  | No   | ...                             | ...                    | ...                  | 7            |
| 25/M/19                  | None                         | Glabella mass        | CT          | No   | Lateral rhinotomy                  | No   | 6                               | Nasal dorsum           | Transverse excision  | 14           |
| 26/M/48                  | None                         | Dorsum mass          | CT          | No   | External rhinoplasty               | No   | ...                             | ...                    | ...                  | 15           |
| 27/M/22                  | None                         | Dorsum mass          | CT          | No   | External rhinoplasty               | No   | ...                             | ...                    | ...                  | 10           |
| 28/M/9                   | None                         | Dorsum mass          | CT          | No   | External rhinoplasty               | No   | 5                               | Nasal tip              | Transverse excision  | 6            |
| 29/M/30                  | None                         | Dorsum sinus         | CT          | Yes  | External rhinoplasty               | No   | ...                             | ...                    | ...                  | 4            |
| 30/M/3                   | Cleft lip and palate         | Nasal tip fullness   | CT          | No   | Transnasal endoscopic              | No   | ...                             | ...                    | ...                  | 5            |
| 31/M/66                  | None                         | Nasal tip fullness   | CT          | No   | Transnasal endoscopic              | No   | 4                               | Nasal tip              | Transverse excision  | 6            |
| 32/M/10                  | None                         | Glabella mass        | CT and MRI  | No   | Medial paracanthal incision        | No   | ...                             | ...                    | ...                  | 1            |
| 33/F/6                   | None                         | Glabella mass        | CT          | No   | Medial paracanthal incision        | No   | ...                             | ...                    | ...                  | 5            |
| 34/F/70                  | None                         | Glabella mass        | None        | ...  | Medial paracanthal incision        | No   | ...                             | ...                    | ...                  | 5            |
| 35/F/9                   | Hemifacial microsomia        | Orbital displacement | CT          | Yes  | Combined intracranial-extracranial | Yes  | ...                             | ...                    | ...                  | 10           |
| 36/M/132                 | None                         | Nasal tip fullness   | MRI         | Yes  | Combined intracranial-extracranial | Yes  | 1                               | Nasal dorsum           | Transverse excision  | 8            |
| 37/M/5                   | None                         | Glabella mass        | CT and MRI  | Yes  | Combined intracranial-extracranial | Yes  | ...                             | ...                    | ...                  | 10           |
| 38/F/132                 | None                         | Glabella mass        | CT          | Yes  | Combined intracranial-extracranial | Yes  | ...                             | ...                    | ...                  | 3            |
| 39/M/17                  | None                         | Widened dorsum       | CT and MRI  | Yes  | Combined intracranial-extracranial | Yes  | ...                             | ...                    | ...                  | 3            |
| 40/F/48                  | None                         | Dorsum sinus         | CT          | Yes  | Combined intracranial-extracranial | Yes  | ...                             | ...                    | ...                  | 6            |
| 41/F/48                  | Craniosynostosis             | Bifid nasal tip      | CT          | Yes  | Combined intracranial-extracranial | Yes  | ...                             | ...                    | ...                  | 8            |
| 42/M/12                  | None                         | Dorsum mass          | CT and MRI  | Yes  | Combined intracranial-extracranial | Yes  | ...                             | ...                    | ...                  | 6            |

Abbreviations: CT, computed tomography; IOIE, intraoperative intracranial extension; MRI, magnetic resonance imaging; POIE, preoperative intracranial extension.

\*Ellipses indicate not applicable.

displacement (n = 1 [2%]). Ten patients (24%) had a history of local infection presenting as erythema and/or intermittent drainage. None had a history of intracranial infection or meningitis. Five patients (12%; 5, 16, 30, 35,

and 41 in the Table) had associated craniofacial abnormalities, including craniosynostosis (n = 1 [2%]), hemifacial microsomia (n = 2 [5%]), bilateral lacrimal duct cyst (n = 1 [2%]), and cleft lip/palate (n = 1 [2%]). None of the

children had a family history of craniofacial or midline nasal abnormality.

Thirty-nine patients (93%) underwent preoperative imaging, including facial radiographs (n=4 [10%]), computed tomography (CT) (n=24 [62%]), magnetic resonance imaging (MRI) (n=4 [10%]), and CT and MRI (n=7 [18%]). The 4 patients (1, 5, 7, and 8) who had plain facial radiographs as the only preoperative workup presented before 1984.

Three patients (7%) had a small subcutaneous nasal mass and did not undergo any preoperative radiographic evaluation; 2 of these (patients 6 and 13) underwent vertical excision and 1 (patient 34) underwent medial paracanthal incision. None of the patients had any intraoperative evidence of a sinus tract or intracranial extension. None of these patients had a recurrence, with a mean follow-up of 7 years (range, 5-10 years). Twenty-eight patients (67%) (Table) did not have clinical or radiographic evidence of intracranial extension. These patients underwent an extracranial approach, including vertical excision (n=15), transverse excision (n=4), lateral rhinotomy (n=2), external rhinoplasty (n=3), transnasal endoscopic (n=2), and medial paracanthal incision (n=2). Twenty-four of these patients did not show any intraoperative evidence of sinus tract or intracranial extension and have done well without recurrence, with a mean follow-up of 8 years (range, 1-15 years). Three patients (patients 25, 28, and 31) presented with a glabellar mass, a dorsum mass, or nasal tip fullness, respectively. We found no evidence of intracranial extension on preoperative imaging studies. All 3 patients underwent resection of the dermoid by means of an extracranial approach, including lateral rhinotomy (patient 25), external rhinoplasty (patient 28), and a transnasal endoscopic approach (patient 31). None of these patients had an intraoperative finding of a sinus tract and/or intracranial extension, yet all 3 had a recurrence in the nasal dorsum (patient 25) or the nasal tip area (patients 28 and 31), with a mean postoperative follow-up period of 5 years (range, 4-6 years). All 3 patients underwent a secondary extranasal approach (transverse excision) of the nasal tip or the nasal dorsum, which revealed evidence of residual dermoid, without any further complication. One patient (16) presented with nasal tip fullness and bilateral lacrimal duct cysts. She underwent CT and MRI, which did not show any evidence of intracranial extension. She underwent intranasal excision of the nasolacrimal cyst and vertical excision of the nasal tip dermoid. No suggestion of a sinus tract was found intraoperatively. However, she presented with a glabellar mass and intracranial extension 2 years after the initial resection. She underwent a frontal craniotomy, and a glabellar mass with intracranial-extradural extension was removed without difficulty. She has done well without any evidence of recurrence, with a follow-up of 2 years after the second procedure.

Three patients (patients 18, 23, and 29) presented with a nasal dorsal sinus or a glabellar mass. Preoperative CT showed no intracranial mass; however, a small sinus tract with a bifid crista galli and/or an enlarged foramen cecum suggestive of possible intracranial extension was noted in all 3 patients. All 3 underwent neurosurgical consultation. The decision was made to proceed with an external nasal approach and craniotomy if in-

traoperative evidence of intracranial extension was found. The external approaches consisted of a vertical excision (patient 18), transverse excision (patient 23), and external rhinoplasty incision (patient 29). All 3 patients had a small fibrous tract extending between and/or along the nasal bone without intracranial extension. The decision was made not to proceed with a craniotomy, and all 3 patients have done well without any evidence of recurrence, with a mean follow-up of 6 years (range, 4-8 years).

Eight patients (19%; patients 35-42) presented with a nasal dermoid with evidence of intracranial extension based on preoperative CT and/or MRI findings. All 8 patients underwent a combined intracranial-extracranial approach, including a subcranial approach (n=3) or a frontal craniotomy with or without external nasal excision (n=5). All 8 showed evidence of intracranial-extradural extension of the dermoid tract. Only 1 patient (36) presented with recurrence in the nasal tip area 1 year after the original resection and underwent transverse excision that revealed scar and epidermal tissue. No other complication was noted, with a mean follow-up of 7 years (range, 3-10 years).

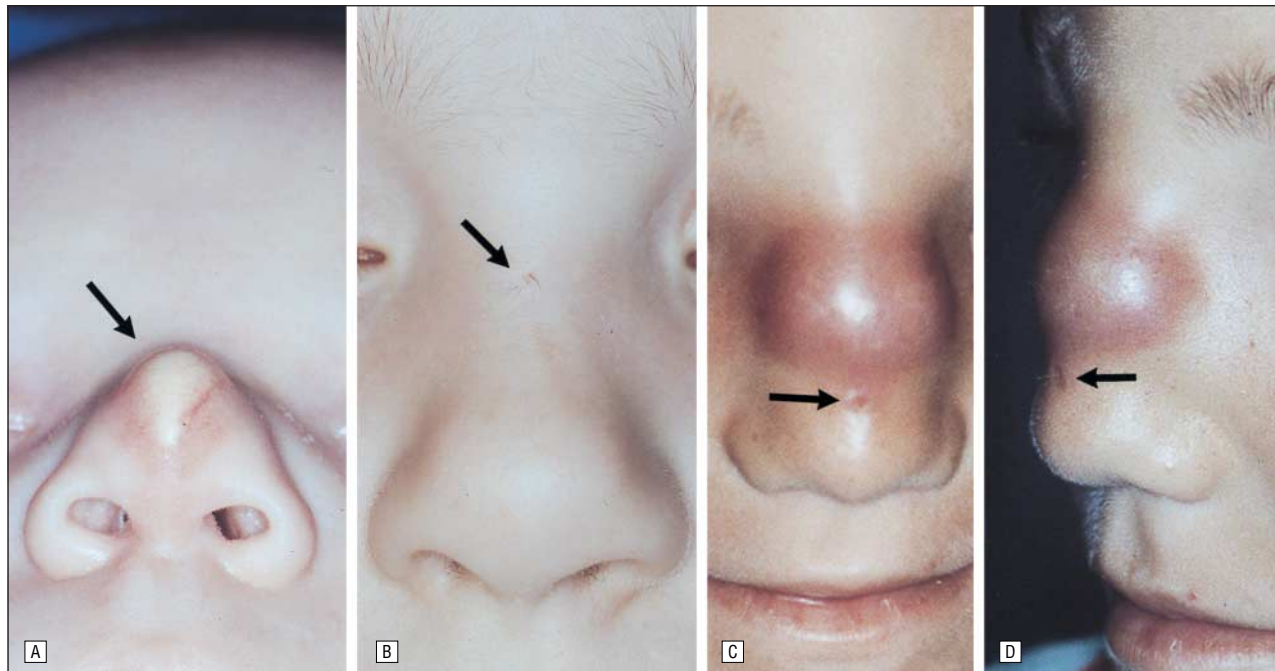
## COMMENT

In 1817, Cruvelier first described a nasal dermoid cyst in a child with a hair-containing sinus opening on the dorsum of the nose.<sup>1</sup> The terminology has been confusing ever since, and various terms such as *dermoid*, *dermoid cyst*, and *dermal cyst* have been used to define the same lesion. Sessions<sup>1</sup> was the first to coin the term *nasal dermal sinus cyst* to include all lesions containing ectoderm (stratified squamous epithelium) and mesoderm (adnexal structure) located in the nose.

## PATHOGENESIS

During embryogenesis, the nose is formed of the following 3 layers: ectoderm, mesoderm, and a deeper layer of cartilaginous capsule. During the eighth and ninth weeks of gestation, the nasal and frontal bones develop by means of intramembranous ossification in the mesoderm but remain separated by the fonticulus nasofrontalis.<sup>5</sup> At this time, a space is formed between the nasal bone and the deeper cartilaginous capsule known as the prenasal space. A small projection of dura extends to the skin. As the nasal process of the frontal bone grows, the skin and dura become separated, and the projection of the dura becomes encircled by the foramen cecum.<sup>5</sup> The dura normally obliterates, thereby severing the neuroectodermal connection. After obliteration of the neuroectodermal connection, the fonticulus nasofrontalis and foramen cecum fuse and the cribriform plates form.

Much has been written about the pathogenesis of the nasal dermoid. In 1893, Bland-Sutton presented the superficial sequestration theory.<sup>1</sup> During the fourth to sixth weeks of embryonic development, the medial nasal processes begin to fuse. According to this theory, epithelial entrapment during the fusion of medial nasal processes could lead to formation of a cyst or a sinus. This theory explains the formation of superficial nasal dermoid, but does not account for a dermoid with intracranial extension.<sup>1,6</sup> In 1961, Littlewood<sup>7</sup> proposed the tri-



**Figure 1.** Presentation of nasal dermoid (arrows). A, Nasal tip; B, hair follicles; C, nasal pit and infected dermoid (frontal view); and D, nasal pit and infected dermoid (lateral view).

laminar theory. Based on this explanation, the formation of nasal cartilaginous tissue is completed by the second month of gestation. The septum is formed of a thin ectodermal layer surrounded by 2 cartilaginous layers. During the third month of gestation, the middle ectodermal layer degenerates, and a sinus or cyst is thought to form due to persistence of the dural-derived ectoderm.<sup>7</sup>

The most widely accepted theory is based on the 1910 theory of Grunwald, later termed the *prenasal theory* by Pratt<sup>2</sup> and the *cranial theory* by Bradley.<sup>8</sup> This theory is based on the finding that as the neuroectodermal tract recedes, dermal attachments can follow its course. As the dura mater recedes from the prenasal space, it may pull the nasal ectoderm upward and inward to form a sinus or a cyst.<sup>2</sup> The resulting epithelial lining forms a dermal sinus or cyst, depending on its connection to the nasal dorsal skin.

### PRESENTATION

Nasal dermoid is the most common congenital midline lesion.<sup>9,10</sup> It constitutes 1% to 3% of all dermoids and 4% to 12% of head and neck dermoids.<sup>11,12</sup> Nasal dermoid is usually seen at birth or in early infancy. The mean age of our patient population was 32 months, with a male predominance, which is consistent with previous reports in the literature.<sup>10</sup> Although familial cases have been reported, we did not document any family history of nasal abnormality in our patients.<sup>1,13,14</sup>

Nasal dermoid typically presents as a midline mass, most commonly along the dorsum, and may be associated with a sinus opening. Intermittent discharge of sebaceous material and recurrent infection is common (**Figure 1**). Hair protruding through a punctum is pathognomonic for nasal dermoid.<sup>15</sup> In our patients, we noted dorsum sinus (n = 10 [24%]), intermittent drainage and/or infection (n = 10 [24%]), and dorsum hair (n = 2 [5%]).

There is no correlation between the initial presentation of nasal dermoid (ie, location or lack or presence of a dorsal ostium) with intracranial extension. Bradley<sup>8</sup> noted that most dermoids were confined to the superficial nasal area (61%), although extension to the nasal cartilage (16%), cribriform plate (12%), nasal bone (10%), and ethmoidal air cells (1%) was noted. Intracranial extension of dermoid has been well reported by Wardinsky et al<sup>15</sup> (10/22 [45%]), Ghestem et al<sup>16</sup> (6/19 [32%]), Bartlett et al<sup>17</sup> (4/9 [44%]), Posnick et al<sup>5</sup> (5/14 [36%]), Sessions<sup>1</sup> (4/13 [31%]), Pensler et al<sup>6</sup> (6/32 [19%]), Denoyelle et al<sup>10</sup> (6/36 [17%]), and Bradley<sup>18</sup> (3/74 [4%]). Intracranial extension of the tract most often passes through the foramen cecum or cribriform plate to the base of the frontal fossa and adheres to the leaves of the falx cerebri extradurally.<sup>15</sup> Intracranial extension with involvement of brain parenchyma has also been reported.<sup>9,19,20</sup> In our series, 8 patients (19%) were documented to have intracranial-extradural extension.

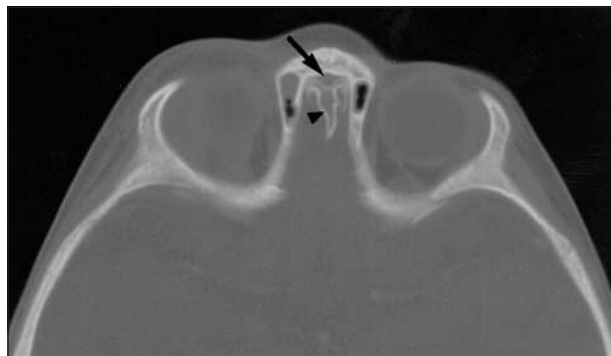
Associated malformative anomalies include aural atresia, deformity of the pinna, mental retardation, hydrocephalus, pharyngeal arch sinus, cleft lip and palate, hypertelorism, and hemifacial microsomia, as noted by Wardinsky et al<sup>15</sup> in 41% of patients, Ghestem et al<sup>16</sup> in 26%, Morgan and Evans<sup>21</sup> in 25%, Sessions<sup>1</sup> in 15%, Posnick et al<sup>5</sup> in 14%, and Denoyette et al<sup>10</sup> in 5.3%. We noted associated craniofacial abnormality in 5 patients (12%), ie, craniosynostosis, hemifacial microsomia, and cleft lip or palate.

### IMAGING

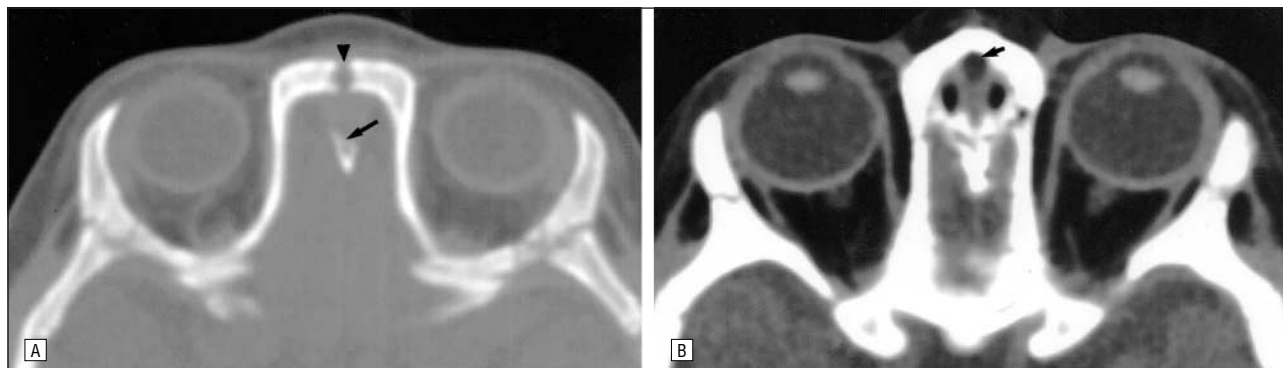
The range of differential diagnosis and the possibility of transcranial extension necessitate a thorough preoperative radiographic evaluation. Imaging modalities include plain radiography, CT, and MRI. McQuown and colleagues<sup>22</sup> described the plain radiographic findings of

a transcranial nasal dermoid that included bony destruction of the glabella, a large infrafrontal-interethmoidal cystic space, widening of the nasal vault, and increased intraorbital distance. The advancement of modern imaging techniques has virtually eliminated the role of plain skull radiography in the evaluation of nasal dermoid.

During the past decade, CT and MRI have become the gold standard in radiographic evaluation of nasal dermoid. Preoperative imaging should assess the anatomy of the sinonasal and cranial base with proper evaluation for intracranial extension. Posnick and colleagues<sup>5</sup> evaluated preoperative CT findings in 14 patients and concluded that patients with intracranial extension showed indication of subcutaneous cyst, bifid crista galli, and enlarged foramen cecum. Using these criteria, many authors have reported false-positive preoperative CT findings in patients with a widened foramen cecum or bifid crista galli who subsequently failed to demonstrate intraoperative evidence of intracranial extension.<sup>1,5,6,10,23</sup> Pensler and colleagues<sup>6</sup> reviewed preoperative CT scans of 32 patients and concluded that an enlarged foramen cecum and bifid crista galli did not correlate with intracranial extension. However, they noted that a normal foramen cecum and crista galli rule out intracranial disease.<sup>6</sup> In our series, 35 patients underwent preoperative CT and/or MRI as part of their workup. Three patients (9%; patients 18, 23, and 29) had a false-positive reading, and 1 patient (3%; patient 16) had a false-negative indication of intracranial extension.



**Figure 2.** Normal anatomy in an axial computed tomographic scan in a 2-year-old child demonstrates the foramen cecum (arrow) ventral to the crista galli (arrowhead).



**Figure 3.** Computed tomographic (CT) scans in a 4-month-old boy with a nasal pit and thickening over the dorsum of the nose. A, Axial CT scan (bone windows) demonstrates an osseous defect between the frontal bones (arrowhead). Asymmetric ossification of the ventral aspect of the crista galli (arrow) is seen, compared with normal anatomy seen in Figure 2. B, Axial CT scan (soft tissue windows) demonstrates a small, rounded, low-attenuation lesion in the expected location of the foramen cecum (arrow). These findings suggest a dermoid extending between the frontal bones to the foramen cecum.

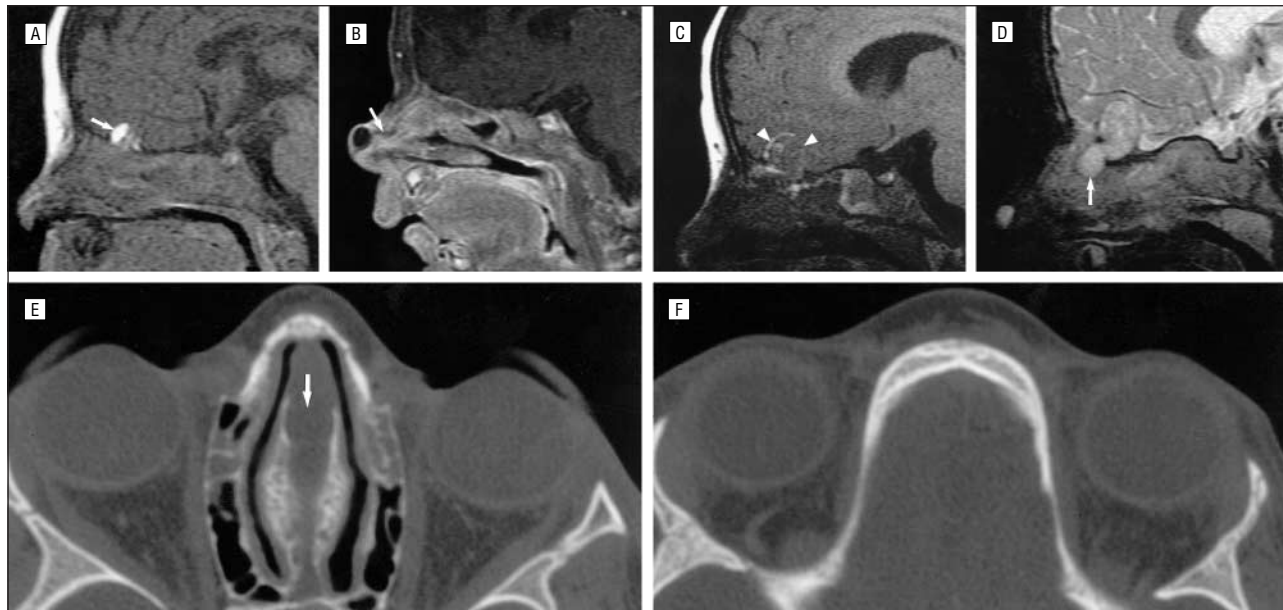
These inaccuracies could be explained by incomplete ossification of the crista galli and foramen cecum in young children.<sup>5,10</sup> Naidich and colleagues<sup>24</sup> reported 14% anatomical variants and incomplete ossification of the crista galli and foramen cecum in children younger than 1 year, which could further confuse the picture and add to the false-positive findings of intracranial extension.

Computed tomography and MRI provide complementary information, and both may be required to adequately delineate bony anatomy and soft tissue characteristics, respectively.<sup>4</sup> Computed tomography (1-3 mm thick, axial and coronal) is optimal for providing bony detail of the anterior or central skull base (**Figure 2** and **Figure 3**). Contrast-enhanced images are recommended to differentiate enhancing cartilage from a skull base defect and to differentiate enhancing nasal mucosa from non-enhancing dermoids. Images should include the entire nasal, ethmoid, and orbital region from the tip of the nose through the anterior cranial fossa. Coronal images should extend posterior to the crista galli. Bony skull base findings include widening of the foramen cecum, ipsilateral osseous defect in the cribriform plate, and a bifid or eroded crista galli.<sup>23</sup>

In the past decade, MRI has become increasingly used for evaluation of nasal dermoid by some investigators.<sup>19,25</sup> Multiplanar (axial, coronal, and sagittal) high-resolution, thin-section MRI should be obtained with T1-weighted images and fat-suppressed T2-weighted or fast spin-echo inversion-recovery pulse sequences. Gadolinium-enhanced, fat-suppressed T1-weighted images are used to depict the anatomy of the enhancing cartilage of the anterior skull base in infants (**Figure 4**). The use of contrast also permits differentiation between nonenhancing dermoids and other enhancing masses such as hemangioma or teratoma. It is important to recognize that in infants, the crista galli is unossified and does not contain marrow fat. As a result, a high-intensity signal on T1-weighted images in the vicinity of the crista galli in the newborn should suggest the presence of intracranial dermoid.

## HISTOLOGY

Light microscopic examination of nasal dermoid reveals a well-defined cyst lined by squamous epithelium of ectodermal origin with adnexal structures (ie, hair fol-



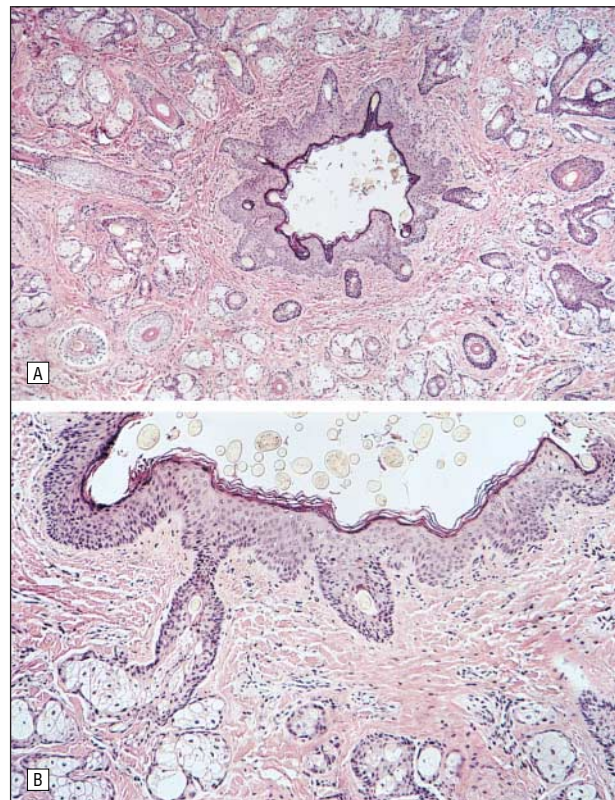
**Figure 4.** This child presented during the neonatal period with a dermoid overlying the tip of the nose. Initial magnetic resonance imaging (MRI) was obtained at 3 months of age. A, Sagittal T1-weighted MRI demonstrates prominent high-intensity signal close to the expected location of the crista galli (arrow). B, Sagittal T1-weighted, fat-suppressed, gadolinium-enhanced MRI reveals the dermoid at the tip of the nose. An apparent thin tract (arrow) leading into the nasal septum was not corroborated on other pulse sequences and imaging planes. The child underwent excision of the nasal tip dermoid, and presented again at 2 years of age with recurrent nasal swelling. C, Sagittal T1-weighted MRI demonstrates loss of the fatty intensity previously ascribed to the crista galli. A rounded, heterogeneous mass (arrowheads) is seen in the midline within the anterior cranial fossa. D, Sagittal T2-weighted MRI reveals recurrence of the dermoid at the tip of the nose and a lobulated dermoid (arrow) within the nasal septum extending cephalad into the anterior cranial fossa. E, Axial computed tomographic (CT) scan demonstrates lobulated remodeling and splaying of the osseous nasal septum by the dermoid (arrow). F, The same CT scan shows lack of the expected ossified crista galli (compared with the normal image in Figure 2).

icles, sebaceous glands, and sweat glands) of mesodermal origin (**Figure 5**). The presence of these adnexal structures distinguishes dermoid from epidermoid cysts, which do not have the same potential for transcranial extension. Unlike teratomas, dermoids do not show any components of endodermal origin.<sup>5</sup>

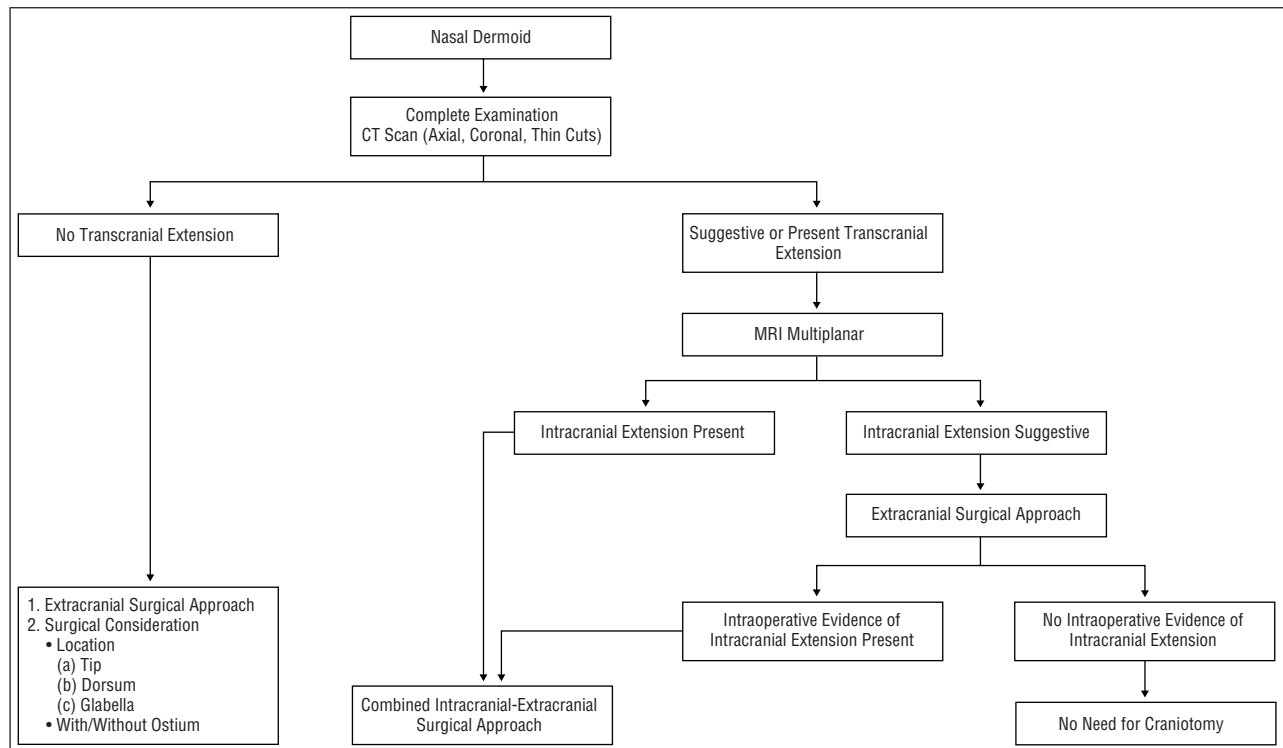
### SURGERY

A thorough preoperative evaluation and appropriate planning are essential before resection of a nasal dermoid. Many different approaches have been advocated for the removal of nasal dermoids in the past 2 decades, ranging from a simple extracranial excision to complex procedures in which intracranial excision and nasal reconstruction are required. Incision and drainage, aspiration, curettage, and subtotal excision fail to eradicate the cyst, resulting in recurrence rates ranging from 30% to 100%.<sup>9,26</sup> A midline vertical incision remains the most common approach.<sup>27</sup> The cutaneous punctum is removed in continuity with the tract and/or the cyst by making an elliptical incision around the sinus opening. Other approaches, such as transverse incision, lateral rhinotomy, external rhinoplasty, inverted-U incision, and degloving procedures, have also been advocated.<sup>5,9,10,11</sup> Posnick et al<sup>9</sup> and Kelly et al<sup>12</sup> reported better cosmetic outcomes with the midline vertical incision compared with lateral rhinotomy and horizontal and inverted-U incisions. However, Bradley<sup>18</sup> reported 37% poor aesthetic results, and Denoyelle et al<sup>10</sup> reported 60% widening of scar associated with vertical incisions.

A number of modified external incisions have been proposed for better cosmetic outcome. External rhino-



**Figure 5.** Histological findings of nasal dermoid (hematoxylin-eosin). A, The sinus tract is lined by keratinizing squamous epithelium. Numerous well-developed pilosebaceous units occupy the sinus wall and open into the lumen. B, The lumen contains numerous cross-sectioned hair shafts. The pilosebaceous units are well-formed and mature.



**Figure 6.** Treatment diagram of nasal dermoid. CT indicates computed tomography; MRI, magnetic resonance imaging.

plasty has been advocated by many authors as the preferred method for extracranial excision with the advantage of better cosmetic results.<sup>5,10,11,28</sup> Weiss et al<sup>4</sup> proposed transnasal endoscopic excision of nasal dermoid when minimal or no cutaneous involvement is found.<sup>4</sup> On the basis of their experience, transnasal endoscopic approach via a bilateral intercartilaginous incision allows excision of the lesion and its tract to the dura of the anterior cranial fossa without the need for a craniotomy. Pollock<sup>29</sup> proposed 2 transverse incisions to remove small to moderate lesions that do not appear to extend intracranially.

When the sinus tract extends deep to the nasal bones, a nasal osteotomy is recommended to improve exposure. The nasal bones are fractured and separated vertically over the dorsum of the nose at the nasofrontal suture. It is essential to follow the tract in its entirety and determine whether there is intracranial extension. When preoperative radiographic and/or intraoperative evidence of intracranial extension is found, a combined approach with a neurosurgical team is safe and effective.

Controversy exists about the surgical strategy when preoperative imaging demonstrates a sinus tract with a bifid and/or widened crista galli but without an intracranial mass. Sessions<sup>1</sup> advocated biopsy of the stalk on the nasal side of the cranial base before craniotomy.<sup>1</sup> He suggested that the tract that appears to extend intracranially often is fibrous and does not need formal excision.<sup>1,6</sup> He proposed that if frozen-section analysis discloses fibrous tissue without the evidence of an epithelial tract, the residual stalk can be suture ligated and the procedure is complete. The need for a craniotomy is thus avoided.<sup>1</sup> This recommendation has been supported by Pensler et al<sup>6</sup> and Bartlett et al.<sup>17</sup> They also proposed an intraoperative serial biopsy of the dermoid stalk at the

cranial base. If no dermal component is found in the dermoid stalk, craniotomy is unnecessary.<sup>6,17</sup>

However, Posnick and colleagues<sup>5</sup> suggested that various epidermal and adnexal elements may appear staggered along the sinus tract at presentation. They postulated that a biopsy at a single site might lead to the false assumption that no dermoid component exists in the more proximal portion. Therefore, these investigators recommend following the dermoid tract to the cranial base via a frontal craniotomy to resect the stalk from the dura.

Our surgical experience at The Children's Hospital, Boston has been large and diverse. Thirty-one patients underwent extracranial excision with no clinical and/or radiographic evidence of intracranial extension. None of these patients showed any evidence of intracranial extension intraoperatively. Three patients (18, 23, and 29) presented with preoperative imaging findings suggestive of intracranial extension of the sinus tract without a definite intracranial component. All 3 patients underwent an extracranial excision only because there was no intraoperative indication of intracranial extension, and biopsy of the tract suggested a fibrous tissue without any dermal component. Eight patients presented with nasal dermoid and intracranial extension, based on preoperative CT and/or MRI findings. They underwent combined intracranial-extracranial excision, which confirmed an intracranial-extradural dermoid in all patients. We have had a total of 5 recurrences (12%) with a mean follow-up of 7.5 years (range, 1-15 years). Four patients (25, 28, 31, and 36) presented with recurrence extracranially, and 1 patient (16), intracranially. The mean time of recurrence was 3.6 years (range, 1-6 years).

Failure to diagnose and resect nasal dermoid properly can result in progressive enlargement, skeletal distor-

tion, infection, meningitis, and intracranial abscess. We advocate a complete evaluation and neurosurgical consultation if any clinical or radiographic indication of intracranial involvement is found. We recommend fine-cut CT with axial and coronal planes through the nose, skull base, and cranium using bone and soft tissue algorithms to determine the extension and possible intracranial involvement. Because of the limitations of CT assessment of soft tissue at the skull base and the reported false-positive findings of scans in the past, complementary multiplanar, high-resolution MRI is strongly recommended (Figure 6).

The timing of resection for an isolated nasal dermoid without intracranial complication remains controversial. We advocate early intervention to prevent the potential risk for infection and the possible need for a more extensive procedure. We agree with Pollack<sup>29</sup> that the surgical approach should fulfill the following 4 criteria: (1) provide excellent access to a midline cyst; (2) allow access to the base of skull; (3) provide adequate exposure for reconstruction of the nasal dorsum; and (4) result in an acceptable scar. An external rhinoplasty approach has a well-concealed scar with an excellent cosmetic outcome. It provides a wide exposure for nasal osteotomy and allows easy access to follow the sinus tract to the skull base. We have used this approach in 3 of our patients without any difficulty. For a lesion in the nasal-glabellar area without a sinus opening that might not be accessible via an external rhinoplasty approach, we prefer a paracanthal incision halfway between the inner canthus and the bridge of the nose or a bicoronal approach. When there is a dorsal ostium, a vertical elliptical incision is essential for removal. However, we agree with Bradley<sup>18</sup> and Denoyelle et al<sup>10</sup> that a poor aesthetic result and widening of the scar can occur after medial vertical excision specifically in the bony dorsal region. We also agree with Sessions,<sup>1</sup> Pensler et al,<sup>6</sup> and Bartlett et al<sup>17</sup> that craniotomy can be avoided if there is a fibrous tract at the cranial base without any evidence, based on intraoperative biopsy findings, of a dermal component. However, it is important to underscore that at present, no study can confirm that the epidermal and adnexal structures extend along the entire sinus tract or are discontinuous as the tract crosses the cranial base. When there is radiographic evidence of intracranial extension, we recommend direct excision by means of a coronal approach, with additional nasal incision if a sinus ostium is present. First, the external cyst or sinus is excised, and then the stalk is followed to the cranial base, followed by a formal craniotomy and removal of the intracranial component (Figure 6).

The recurrence rate of nasal dermoid is low, but may occur several years after the initial surgery. Therefore, the long-term follow-up of all patients with a history of nasal dermoid is essential.

## CONCLUSIONS

Nasal dermoid is a rare congenital lesion, which often poses diagnostic and surgical dilemmas. Preoperative evaluation is essential to rule out intracranial extension. Workup should include fine-cut CT, and complementary MRI should be considered if there is concern for intracranial extension. Surgical management is dependent on the lo-

cation and extent of the lesion, ranging from local excision to a combined intracranial-extracranial approach.

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