



Incidental Findings on Brain and Spine Imaging in Children

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abstract

In recent years, the utilization of diagnostic imaging of the brain and spine in children has increased dramatically, leading to a corresponding increase in the detection of incidental findings of the central nervous system. Patients with unexpected findings on imaging are often referred for subspecialty evaluation. Even with rational use of diagnostic imaging and subspecialty consultation, the diagnostic process will always generate unexpected findings that must be explained and managed. Familiarity with the most common findings that are discovered incidentally on diagnostic imaging of the brain and spine will assist the pediatrician in providing counseling to families and in making recommendations in conjunction with a neurosurgeon, when needed, regarding additional treatments and prognosis.

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INTRODUCTION

The importance of brain imaging in contemporary medical practice can hardly be exaggerated, and technological advances have been matched by expansion of access to this technology. When the senior author entered medical school, there was precisely 1 new first-generation computed tomography (CT) unit at his prestigious referral teaching hospital. Now, much more sophisticated devices are found even in small facilities. In recent years, the use of diagnostic imaging of the brain has expanded dramatically, and requests for consultation to assess the meaning of unexpected findings have multiplied. Although insurance carriers scrutinize them, few population-based data have been published describing actual numbers of studies performed, particularly among children. In every year from 2008 through 2012 in the state of Delaware, slightly more than 2% of all children enrolled in Medicaid underwent CT scanning or MRI of the brain (P. White, Delaware Division of Medicaid and Medical Assistance, personal communication, 2013). A recent review of 6 large integrated health systems in the United States found that MRI use quadrupled between 1996 and 2010, reflecting a 10% annual growth rate.¹ MRI use is influenced by many factors, including increasing availability, patient-generated demand, and “defensive” practice.² Although rates of MRI use vary substantially by geographic region within

the United States,^{3,4} overall MRI use is substantially higher in the United States than in most other industrialized countries. In the most recent data from the Organization for Economic Cooperation and Development, MRI use in the United States per 1000 population was 97.7, the highest rate in the survey and more than double the Organization for Economic Cooperation and Development average.⁵ As a result, the increase in incidental brain and spine findings is of particular importance in the United States. An unintended consequence of such widespread diagnostic imaging is the discovery of many incidental findings unrelated to the original reasons for the studies.

BENIGN ENLARGEMENT OF THE SUBARACHNOID SPACES

Benign enlargement of the subarachnoid spaces (BESS), also known as “benign external hydrocephalus,” is a transient developmental phenomenon that is really a variant of normal and is commonly seen in neurosurgery practice.^{6–8} Because the indication for the brain imaging that discloses BESS is usually macrocephaly, it may not be an incidental finding in a strict sense, but it is incidental inasmuch as treatment is seldom necessary. In the era of CT scanning, it was the focus of some confusion, the term “benign subdural effusions of infancy” suggests, but contemporary ultrasonography with Doppler or MRI can make reliable distinctions between subdural fluid collections and prominence of the subarachnoid spaces. Likewise, the term “external hydrocephalus” suggests a disturbance of cerebrospinal fluid (CSF) physiology, which has never been substantiated experimentally.

The clinical picture is fairly consistent. Typically, the affected patient exhibits accelerated head growth in midinfancy but otherwise thrives. Often, 1 parent has a head circumference at or beyond the 95th

percentile. Despite the presence of macrocephaly, the fontanel is slack and there is no suture separation. Brain imaging may show a minor degree of ventriculomegaly that is not in proportion to the expansion of the subarachnoid spaces. Although seldom documented because of its benign clinical course, the natural history of BESS is resolution later in childhood; and from a clinical standpoint, the child’s head circumference can be expected to drift gradually back toward the top of the normal range over a period of years.⁹ The developmental prognosis of BESS is the topic of some discussion in the literature and remains imprecisely defined, even after imaging mimickers, such as achondroplasia, Sotos syndrome, and mucopolysaccharidoses, have been excluded.^{8,10,11} Generally, after initial gross motor delays attributable to the excessive size of the head, development is normal, but careful observation is warranted.

Neurosurgical referral is not necessary in the absence of ventricular enlargement or subdural fluid collections unless parents require the reassurance of the subspecialist. Although a degree of ventricular enlargement may accompany BESS, a report of ventricular enlargement raises a question of hydrocephalus that must be addressed by a neurosurgeon. Likewise, a description of subdural hygroma or chronic subdural hematoma indicates referral.^{6,12–14} In addition, if the interpretation of the initial imaging study fails to distinguish between BESS and subdural hematoma or hygroma, neurosurgical referral is indicated necessarily as well. The disproportion between the volume of the cranium and the volume of the brain that characterizes BESS is widely believed to create susceptibility for development of subdural fluid collections, but the finding of a subdural collection cannot be dismissed. A recent cohort

study identified subdural collections in 4 of 177 young children with BESS (2.3%), 1 of whom was determined to be a victim of abuse.¹⁵ Even in the setting of BESS, chronic subdural hematoma still constitutes an indication for investigation of the possibility of abuse, including dilated fundoscopic examination and radiographic skeletal survey.¹⁵

CHOROID PLEXUS CYST

Choroid plexus cysts are common findings on antenatal ultrasonography in the second trimester. Prevalence rates between 0.6% and 2.3% have been reported.^{16–19} Choroid plexus cysts are of some significance in perinatal medicine because of their association with fetal aneuploidy in the setting of other risk factors. Although Shuangshoti and Netsky²⁰ reported small choroid plexus cysts in the majority of unselected autopsy specimens, they are infrequent incidental findings on brain ultrasonography in infancy and are seldom noted on brain imaging studies in older children and adults. Thus, choroid plexus cysts in the fetus are generally believed to regress before or shortly after birth. The neurosurgical literature contains many case reports of choroid plexus cysts that were symptomatic at presentation from obstructive hydrocephalus, but we are aware of only a single example in print of a cyst detected in the second trimester that persisted to term and progressed postnatally to become a clinical problem.²¹ Thus, there seems to be little cause for concern about postnatal neurosurgical complications of antenatally detected choroid plexus cysts. Indeed, the authors are unable to reference any published guidelines for follow-up imaging in infancy. Incidentally detected choroid plexus cysts associated with mass effect or hydrocephalus certainly require neurosurgical referral. There is no

evidentiary basis for recommendations about smaller cysts, but neurosurgical consultation is likely to prove more conclusive and less expensive than sequential imaging studies.

CHOROIDAL FISSURE CYST

The choroidal fissure can be found on the mesial surface of the temporal lobe between the hippocampus and the diencephalon, and a choroidal fissure cyst is a loculated cavity filled with CSF lying in the fissure. The relationship of the cyst to the fissure is best visualized on coronal brain imaging; on axial imaging (as in most CT scans), the location of this structure can be misconstrued.²² Almost without exception, choroidal fissure cysts are incidental findings.²² The natural history of choroidal fissure cysts has not been documented in detail, but in general, cysts without mass effect in school-aged children are static and require no follow-up.^{23,24} In infancy, choroidal fissure cysts can exhibit an unstable, progressive course.²⁵ Because the imaging diagnosis can be subtle and because other cystic lesions of the mesial temporal structures can have very different clinical implications, neurosurgical referral is indicated.

LIPOMA OF THE FILUM TERMINALE

Filum terminale lipomas are a type of lumbosacral lipoma in which fat is entirely within the filum terminale and separate from the conus medullaris. Filum lipoma is sometimes detected as a genuinely incidental finding but more often is a finding of uncertain significance that comes to light in the course of an investigation of intractable urinary incontinence, chronic constipation, pes cavus, gait abnormalities, or anomalies of the intergluteal crease. The prevalence of filum lipoma at autopsy has been reported to be 6%.²⁶ Its prevalence among children and adults undergoing MRI for

unrelated reasons is 1.5% to 4%.²⁷⁻³¹ The concern is that filum lipoma may be a cause of or marker for spinal cord tethering.³² Symptoms of tethered cord syndrome include back and lower extremity pain, urologic abnormalities, lower extremity weakness, gait disturbance, and foot and ankle deformities such as pes cavus.³³⁻³⁵ With the increasing use of MRI, filum lipomas have been identified more frequently in asymptomatic individuals.^{28,36,37} The clinical significance of a filum lipoma in an asymptomatic child is a subject of debate.^{27,36,37} Division of a lipomatous filum for relief of tethering (a relatively simple neurosurgical procedure) is undertaken commonly in selected patients with pain, progressive neurologic deficits, scoliosis, or disturbances of bowel and bladder function.^{34,38,39} Unfortunately, symptoms such as back pain and urologic complaints are common in children without neurologic abnormalities, and many of these symptoms resolve with time, medical management, and behavioral therapy.^{40,41} Therefore, the management of children with subjective symptoms of tethered cord syndrome is controversial, especially for those patients without lower than normal position of the spinal cord on imaging. The untreated natural history of asymptomatic filum lipoma is probably benign for most patients.²⁹ For this reason, prophylactic untethering is not indicated for most patients, although some authorities in the past have promoted aggressive prophylactic treatment.³⁸ Neurosurgical evaluation is appropriate, but surgical intervention will seldom prove necessary for incidentally discovered lesions.

PERINEURAL (TARLOV) CYSTS

Perineural cysts arise from the spinal nerve root or dorsal root ganglion. They have a meningeal lining and

contain CSF that is in variable communication with CSF of the thecal sac, and nerve rootlets and ganglion cells lie in the cyst or within the cyst wall. The eponym "Tarlov" is applied to cysts arising in the sacral region, which is by far the most common site.⁴² Among adults, the imaging prevalence of Tarlov cysts has been reported to be between 1.5% and 4.6%.^{31,43-45} The prevalence among children is much lower, consistent with gradual acquisition and expansion of the lesions based on upright posture and hydrostatic CSF pressure. Incidental cysts greatly outnumber symptomatic ones. Clinical assessment must focus on correlating anatomy with symptoms and with the segmental neurologic examination. Perineural cysts are a doubtful explanation for nonspecific low back pain, but they can cause radicular pain and neurologic signs. In the sacral region, perineal pain, bladder symptoms, and sexual dysfunction are relevant. Characteristic features are exacerbation by Valsalva-type events and relief with recumbency. Anatomically consistent radiculopathy indicates neurosurgical referral, and when the nature of the presenting symptoms is obscure, neurosurgical consultation is necessary as well. No imaging surveillance is required in most cases.

INCIDENTAL BRAIN TUMORS

There is no alternative to neurosurgical referral and management of a child with an imaging study interpreted to show an incidental brain tumor. Nevertheless, the primary care physician can address family anxiety and adjust expectations more effectively with some understanding of management issues.

The great majority of incidentally discovered brain tumors in childhood are benign. This fact is readily explained by the overall slight predominance of benign over

malignant tumors in childhood and the much longer time interval between the threshold of imaging detection and production of symptoms for benign tumors.⁴⁶ Examples of benign childhood brain tumors include astrocytoma (World Health Organization grades 1 and 2), ganglioglioma, and dysembryoplastic neuroepithelial tumor.

Reliable statistics describing the prevalence of asymptomatic brain tumors in childhood do not exist. Imaging case series of children with headache are uninformative for a variety of reasons. Such series feature only highly selected patients. Brain tumors reported in these series are seldom explicitly stated to be incidental, and no accounting is made for possible indolent tumors among what are called “white matter lesions” or “parenchymal tissue abnormalities.”⁴⁷⁻⁵¹ The recruitment of normal adults for MRI research is generating a growing volume of literature on the prevalence of incidental findings among genuinely asymptomatic individuals, but there is very little information on normal pediatric research subjects. Kim et al⁵² found 1 likely cerebellar tumor among 225 children (0.4%) enrolled in MRI research. Jordan et al⁵³ found 4 asymptomatic brain tumors among 953 children (0.4%) with sickle cell disease recruited to an imaging study of silent cerebral infarction. Seki et al⁵⁴ noted no brain tumors among the MRI studies in 150 normal Japanese children between 5 and 8 years of age. No brain tumors were recognized among 96 normal children from Malawi studied via MRI by Potchen et al.⁵⁵ Because the neoplastic nature of an isolated incidental finding can often be confirmed only by growth revealed on sequential studies, cross-sectional surveys of normal subjects necessarily produces low estimates. Whatever their true prevalence may be, incidental brain tumors are a regular feature of pediatric

neurosurgery and neurooncology practice.⁵⁶⁻⁵⁸ As expected, benign lesions predominate. In 1 relatively large series, only 3 of 47 tumors were malignant.⁵⁸ Management is individualized on the basis of the appearance and location of the lesion. Lesions recognized to be malignant, such as medulloblastoma, are investigated and treated aggressively. Lesions for which even a small degree of expansion may magnify the difficulty of surgical treatment, such as craniopharyngioma, are also investigated and treated expediently. On the other hand, the neurosurgeon may choose to observe lesions that appear to be benign glial tumors.⁵⁶⁻⁵⁸ Benign glial tumors in childhood have a favorable natural history compared with histologically similar lesions among adults, for whom eventual and lethal malignant degeneration is the rule. Benign childhood gliomas grow slowly, and incidental lesions can be treated surgically for cure before they cause symptoms. Malignant degeneration is rare among children, so withholding of treatment until imaging surveillance reveals growth is often a safe and attractive strategy. The emotional stress of periodic imaging is burdensome for some families, so patient and family preferences must be weighed.

INCIDENTAL VASCULAR LESIONS

Four types of vascular malformations are recognized in the central nervous system: arteriovenous malformation (AVM), cavernous hemangioma, developmental venous malformation (DVM), and telangiectasia. The last of these lesions, telangiectasia, is almost invisible on CT scan. It requires selected MRI sequences, contrast administration, and ideally, high magnet strengths for clear identification.⁵⁹ It is believed to pose no risk of hemorrhage and is not a common incidental finding in childhood.⁶⁰

AVMs vary greatly in size and complexity, but the defining feature is

the direct connection between the arterial and venous systems without an intervening capillary bed. AVMs pose a lifelong risk of hemorrhage at arterial pressures, leading to high rates of neurologic disability and possible mortality. Incidental discovery of a cerebral AVM is an indication for neurosurgical referral, and families may be counseled that a recommendation for proactive treatment may be forthcoming.

Cavernous hemangioma, or “cavernoma,” is a less-threatening lesion composed of thin-walled vessels of varying luminal size without any intercalated normal brain tissue. Constituent vessels that are not thrombotic convey blood at capillary or venous pressures and at very low flow rates. Cavernomas are invisible on catheter angiography, but they have a vivid and distinctive MRI appearance. Multiple lesions are not uncommon, and follow-up may reveal enlargement of existing lesions and de novo appearance of new ones. The classic autopsy prevalence is 0.4%.⁶¹ The imaging prevalence of incidental cavernomas in childhood has been estimated at 0.3%, and prevalence seems to increase with age.⁶² Cavernomas become symptomatic either with seizures or from hemorrhage. They are distinct from AVMs in that hemorrhages are smaller, are less likely to cause disability depending on anatomic location, and are infrequently fatal. The annual risk of hemorrhage is lower as well; a recent report of a pediatric series estimated a 0.2% per lesion per year rate for incidental cavernomas.⁶² Finally, in comparison with AVMs, the surgical management of cavernomas is relatively straightforward. Neurosurgeons are generally reluctant to operate on asymptomatic cavernomas, but exceptions may be made on the basis of such factors as interval enlargement, anatomic location, or family preference. The referring physician may reassure the family that an incidental cavernoma poses

no immediate threat to the life and well-being of the child, but allowance must be made for exercise of the consultant's judgment in treatment recommendations.

What were formerly called "venous angiomas" are now denominated "developmental venous malformations" (DVMs), because they have come to be recognized as nothing more than anomalous patterns of venous drainage of normal brain parenchyma. DVMs are common incidental imaging findings with an autopsy prevalence of 2.5%.⁶¹ They can be seen at any level of the neuraxis, but in the cerebral hemispheres they have a typical morphology: a dominant, radially oriented vein drains superficially or deeply and converging on the origin of this vein are a number of smaller tributaries. This typical morphology, the "caput medusae," recalls the appearance of snakes emanating from the head of Medusa in Greek mythology. DVMs can be seen in association with other vascular malformations, most commonly cavernomas, in which case clinical management is dictated by the character of the associated lesion.⁶³⁻⁶⁵ In isolation, DVMs have no recognized causal relationship with hemorrhage, seizures, or any other clinical phenomena.⁶⁴ Children with incidentally discovered DVMs deserve neurosurgical review, but in the absence of any associated vascular pathology, isolated DVMs do not require treatment or follow-up.^{60,65}

PITUITARY ABNORMALITIES

Unanticipated imaging findings involving the pituitary gland are so common that the term "pituitary incidentaloma" has established itself in the literature.⁶⁶ The autopsy prevalence of pituitary tumors has been estimated at 14%,⁶⁷ but the mix of incidental findings also includes cysts of various kinds, physiologic and pathophysiologic hypertrophy of

the gland, the so-called empty sella syndrome, and morphologic anomalies of the sella itself that can distort the appearance of the gland.⁶⁸⁻⁷¹

As is true of incidentally discovered brain tumors, there is no alternative to neurosurgical referral for incidentally discovered pituitary lesions, but the general pediatrician can set expectations and initiate the investigation. A sensible first step is to obtain a menstrual history, if appropriate, and screening endocrine data: cortisol, thyroxine (T₄), triiodothyronine (T₃), thyroid-stimulating hormone, insulin-like growth factor 1 or somatomedin, prolactin, and, in postmenarchial girls, β -human chorionic gonadotropin. In this context, note must be made of the natural hypertrophy of the pituitary gland associated with pregnancy⁷⁰ and the unnatural but secondary hypertrophy of the pituitary gland associated with primary hypothyroidism.⁷¹ Unlike in adults, the most common secretory tumor of the pituitary gland in childhood is the corticotroph adenoma, followed by prolactinoma and somatotroph adenoma, but management proceeds according to the same principles as in adulthood.⁷² Visual field testing is indicated for expansive lesions large enough to distort the optic chiasm, and as for adults, surgical intervention is usually indicated.

Normal developmental expansion of the pituitary gland in peripubertal girls continues to be an occasion for confusion.⁷³ Normative values for the dimensions of the gland have been analyzed.⁷⁴ The gland may take on a spherical shape, the diaphragm of the sella may assume a convex contour, and the gland may abut the optic chiasm. Uniform signal intensity, prompt homogeneous contrast enhancement, and normal endocrine laboratory data confirm the physiologic nature of these changes. Similar but less pronounced features

may be seen in boys.⁷⁴ Neurosurgical referral may be considered in doubtful cases, but experienced neurosurgeons are very reluctant to call an enlarged pituitary abnormal in this clinical setting.⁷³

Patients with other incidental findings and normal endocrine laboratory values will be referred for neurosurgical consultation as well, but families may be reassured that imaging surveillance will likely be the recommendation. The frequency and duration of imaging surveillance remain a matter for neurosurgical judgment.⁷⁵

ARACHNOID CYSTS

Arachnoid cysts are very common. Most recent large studies have estimated arachnoid cyst prevalence on imaging at ~2%.⁷⁶⁻⁷⁹ The prevalence of arachnoid cysts does not change significantly with advancing age. Boys are nearly twice as likely to harbor arachnoid cysts as girls.^{77,80-83} Most arachnoid cysts are found in the anterior middle fossa and retrocerebellar locations.^{76,77,80-83} Middle fossa arachnoid cysts have a left-sided predominance.^{76,77,79,80,83} Arachnoid cysts may be very large or very small, but size does not correlate precisely with symptoms or the need for treatment.

Arachnoid cysts occasionally present with neurologic signs or symptoms; however, in most cases they are asymptomatic and found incidentally.^{77,84,85} Because arachnoid cysts are common incidental findings, individuals frequently present with both an arachnoid cyst and an unrelated condition or symptom. Clinicians should exercise caution when ascribing any nonspecific symptom, such as headache, behavior disturbance, or epilepsy, to the presence of an arachnoid cyst.^{85,86} Furthermore, such symptoms often persist after surgical treatment of an

arachnoid cyst.^{81,86} Although arachnoid cysts may occasionally enlarge or decrease in size, most do not change substantially over time.^{76,77}

Although most arachnoid cysts should not be treated, there are certain clear exceptions that will benefit from treatment. Cysts should be treated in most cases if they are causing clear and specific neurologic symptoms.⁸⁷⁻⁹⁰ Even small cysts may require treatment if they block normal CSF pathways and cause symptomatic hydrocephalus.

Arachnoid cysts in the suprasellar location are especially likely to cause symptoms and require treatment. As with any surgical procedure, these operations are associated with potential morbidity,^{81,91-93} and the decision to treat surgically should be made very carefully and only after taking the prevalence and natural history of these cysts into account. Most middle fossa and retrocerebellar arachnoid cysts should not be treated. A minority of neurosurgeons continue to make an argument that arachnoid cysts may alter cognition and have used this argument as a justification for surgery in some cases.^{94,95} Evidence of mass effect on imaging is not, by itself, a sufficient indication for surgical treatment of an arachnoid cyst. Any large intracranial cyst can have the appearance of mass effect on imaging. This criterion, therefore, is too inclusive to be used as a reliable indicator for selecting patients for surgical treatment. Because arachnoid cysts are common and the untreated natural history of most arachnoid cysts is benign, most believe that surgical treatment should be avoided except in the unusual instance in which a cyst is clearly responsible for specific symptoms.

Arachnoid cysts may occasionally develop associated subdural hygromas resulting from a spontaneous or traumatic tear in the outer cyst lining.^{86,96,97} These hygromas are rare, and although they

are often symptomatic, they do not always require surgical treatment. Furthermore, surgical treatment of arachnoid cysts can cause iatrogenic hygromas.^{86,87,93} For these reasons, prophylaxis against future hygroma risk should not be regarded as an adequate indication for surgical treatment. Hemorrhage may occasionally occur into an arachnoid cyst after trauma.⁹⁷⁻¹⁰⁰ Because this is also a very rare event, prophylaxis against future hemorrhage risk should not be used to justify surgical treatment in an asymptomatic child. Furthermore, hemorrhages associated with arachnoid cysts are associated with generally good outcomes, even when surgical evacuation of the hemorrhage is deemed necessary.⁹⁷

PINEAL CYSTS

Pineal cysts are also frequently discovered on brain MRIs in children.¹⁰¹⁻¹⁰⁶ Pineal cyst prevalence changes with age, but these cysts are found more frequently in girls than in boys in all age groups.^{101-103,105-109} They are relatively unusual in infants, becoming more common in childhood and adolescence, and peak in young adulthood when they are seen in as many as 2% to 4% of brain MRIs. They become increasingly uncommon with advancing age in adulthood.^{101,103,105,110} Because most pineal cysts are too small to be detected on MRI, the prevalence of microcysts on autopsy studies is even higher.^{111,112} Despite the frequent occurrence of this finding on imaging, the discovery of a pineal cyst often results in a neurosurgical consultation.^{105,107,113}

Pineal cysts have a typical appearance on MRI. Cyst contents may be either isointense or slightly hyperintense to CSF on T1-weighted and hyperintense on T2-weighted imaging.^{104,108,114-116} The cyst rim appears smooth and thin in most cases, but multiple septations within

the cyst itself is a common finding.^{111,112,117} Many benign pineal cysts show evidence of irregular nodular enhancement or ring enhancement on MRI^{118,119} because of surrounding venous structures or the displaced pineal tissue.^{101,103,120,121}

Several groups have reported on the natural history of untreated pineal cysts over time.^{101-103,105,107,120} Although some cysts enlarge and others involute, most cysts remain the same size over several years of follow-up. Importantly, the vast majority of asymptomatic cysts remain asymptomatic. Some growth over time is more often seen in children, and involution is more often seen in adults. Taken together with the cyst prevalence data, the natural history data suggest that pineal cysts frequently arise and change during childhood and then involute during adulthood. For this reason, cyst growth alone in an asymptomatic child should be regarded as the natural course of these cysts and should almost never, by itself, be used as a justification for surgery.¹⁰¹⁻¹⁰³

Pineal cysts are almost always asymptomatic. Rarely, hydrocephalus may result from cerebral aqueductal obstruction due to a large pineal cyst.¹⁰⁶ Some reports have suggested that cysts larger than 1 cm in maximal dimension are more likely to be symptomatic.^{106,112,119,122} Most cysts that cause hydrocephalus are greater than 2 cm in maximal dimension.¹²³ Rarely, large pineal cysts have also been associated with gaze palsy and Parinaud syndrome.^{106,108,118,122,124,125} Although large cysts may be symptomatic, this occurrence is rare. The vast majority of even large cysts may be expected to present incidentally and remain asymptomatic.^{101-103,107,115,120} Given their common incidence on imaging, pineal cysts have a frequent coincidental association with common symptoms such as headache.

A substantial number of children present to their pediatrician with headaches and a pineal cyst that is coincidentally rather than causally associated. Although there are reports of pineal cyst surgery performed for the treatment of chronic headaches,^{119,122,126,127} most surgeons do not regard a history of chronic headache in the absence of hydrocephalus as an indication for surgical treatment of a pineal cyst. Not surprisingly, headaches can be expected to have reliable postoperative symptomatic relief only if the headaches were caused by hydrocephalus.¹²³

Pineal cysts are usually incidentally identified and, once identified, do not result in clinical symptoms in most cases. Specialty consultation and follow-up imaging should be considered merely optional for most asymptomatic children with a conclusive imaging diagnosis of a pineal cyst. In almost every case, cysts smaller than 1 cm will not require neurosurgical evaluation. Larger cysts may also be managed without surgery in many cases but may benefit from an evaluation by a neurosurgeon.

CHIARI AND SYRINGOMYELIA

Chiari malformation type I (CM) is a condition found frequently in children as well as in younger and middle-aged adults. CM is occasionally associated with neurologic symptoms. The classic presentation of CM is headache precipitated by Valsalva-type events, such as sneezing, coughing, or straining. Typically, these headaches are short-lived and located posteriorly. Other symptoms are legion but often nonspecific. These include visual disturbances, dysphonia, dysphagia, sleep apnea, clumsiness and incoordination, and sensory disturbances.^{128,129} CM may also lead to the development of syringomyelia, which, in turn, may lead to other symptoms including

motor and sensory difficulties, pain, and scoliosis.¹³⁰⁻¹³⁴ Surgical case series tend to overestimate the frequency of syringomyelia in patients with CM.¹³⁵⁻¹³⁷ The prevalence of syringomyelia in the setting of CM estimated from imaging databases has been reported to be between 12% and 23%.^{138,139} Syringes are less likely to be found associated with CM in children younger than 5 years but may develop later in childhood.¹³⁹ Lower position of the cerebellar tonsils in CM is associated with a greater likelihood of syringomyelia.¹³⁹⁻¹⁴¹

It is probable that Chiari symptoms and the formation of spinal syringes are the result of crowding at the foramen magnum that leads to abnormal movement of CSF at the craniocervical junction.¹³⁴ Children with CM often have a smaller than normal posterior fossa volume, resulting in crowding of the posterior fossa and foramen magnum contents.¹⁴²⁻¹⁴⁴ This crowding at the foramen magnum may be appreciated on sagittal MRI by the typical “peg-shaped” appearance of the cerebellar tonsils. Because “crowding” is difficult to quantify or objectively determine, the diagnosis of CM on imaging is usually made by a determination of cerebellar tonsil position on MRI. Most often, children are assigned a diagnosis of CM if the cerebellar tonsils are found to be 5 mm or more below the foramen magnum, usually defined as a line between the basion and opisthion in the midsagittal plane.^{78,139,145-148} This definition of CM resulted from the publication of several clinical studies involving small numbers of patients at the dawn of the MRI era.^{145,146} Although cerebellar tonsil measurements that define CM on MRI may be a convenient marker for crowding at the foramen magnum, the correlation is not exact. Patients with less than 5 mm of descent can occasionally present with typical CM symptoms or even syringomyelia attributable to crowding at the

foramen magnum.¹⁴⁹ Conversely, many patients with cerebellar tonsils that are more than 5 mm below the foramen magnum are completely asymptomatic.^{139,150,151} Although CM does present with symptoms in many cases, it is also a very common incidental finding in asymptomatic individuals. Unfortunately, the current clinical tendency appears to be unreservedly admitting such patients to a patho-anatomic group we call CM on the basis of tonsil position alone.

Like most morphometric measurements, cerebellar tonsil position with respect to the foramen magnum follows an essentially normal distribution in all age groups.¹⁵² Those on the lower end of this population distribution fall within the range that may be diagnosed as CM on MRI. The average position of the cerebellar tonsils trends inferiorly during childhood and young adulthood, but the trend reverses with advancing age in adulthood.^{147,152} Females have, on average, a lower tonsil position than males, and CM is more frequently diagnosed in females.^{139,140,151}

If the current imaging definition of tonsils 5 mm below the foramen magnum is used, CM is not rare.^{78,138-140,153,154} CM is found in approximately 0.8% to 1% of all patients undergoing MRI when age is not considered.^{78,138,153} When prevalence is stratified by age, it is clear that the prevalence in children and young adults is greater.¹⁵² As many as 3.6% of children undergoing MRI of the brain or cervical spine have CM by imaging criteria; most children who meet the definition of CM by imaging criteria are asymptomatic.^{139,154}

There are no universally accepted surgical indications for CM. Asymptomatic individuals without a spinal syrinx are only exceptionally considered for surgical treatment. Those with clear symptoms who also have syringomyelia are generally

considered excellent candidates for Chiari decompression surgery.¹⁵⁵ Treatment of those with nonspecific symptoms such as headache (without the usual characteristic features of Chiari-associated headache) is controversial. Because headaches are a common symptom in the general population and CM is a common incidental finding on imaging, surgeons must exercise restraint when selecting these patients for treatment. The natural history of an incidentally discovered Chiari may be expected to follow a benign course in most cases.^{150,151,156} Symptoms and MRI findings are stable over time for most patients, with no symptoms or minimal symptoms that are followed without surgical treatment, although spontaneous improvement or worsening of both the CM as well as syringomyelia does occasionally occur.^{150,151,156}

Although most cases of clinically relevant syringomyelia are associated with CM, it may also be seen in patients with spinal cord tumor, tethered cord, or arachnoiditis.^{157,158} In general, the management of syringomyelia should be directed at the primary disorder. When no primary disorder is identified, the syrinx is considered idiopathic. The untreated natural history of idiopathic syringomyelia is excellent,^{158,159} and the majority of such cases should not be considered for surgical treatment. Small spinal syringes may be difficult to distinguish from minimal dilations of the central canal of the spinal cord. Central spinal cord fluid collections of less than 2 or 3 mm in maximal diameter on axial imaging generally represent only a dilated central canal and are not associated with symptoms.

Children with imaging diagnoses of CM or syringomyelia generally require neurosurgical consultation, but the pediatrician can reassure parents that incidental, asymptomatic

findings are unlikely to require surgical treatment.

CONCLUSIONS

Unexpected findings on imaging studies now account for a large fraction of new patients referred to pediatric neurosurgical practices. They are the cause of a great deal of parental distress, and they put patients at risk of unnecessary additional testing and unnecessary surgery. Rational use of diagnostic technology and subspecialty consultation can minimize these confusing family experiences, but the diagnostic process will always generate an irreducible minimum of unexpected findings that must be accepted and managed for the patient's welfare. Familiarity with the most common entities allows the pediatrician to allay parental anxieties with informed preliminary counseling and to set appropriate priorities for subsequent referrals and investigations. By improving their knowledge base about incidental findings on neuroimaging, pediatricians can provide guidance to families, with neurosurgical consultation when needed regarding clinical relevance, need for additional testing, and need for follow-up.

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