The cutaneous stigmata of occult spinal dysraphism (OSD) are well known to most neurosurgical practitioners: the patch of hair, the subcutaneous lipoma, the dermal sinus tract, the lumbosacral dimple, and more exotic signs, such as the caudal appendage and the so-called “cigarette burn” lesion. These stigmata are a matter of neurosurgical lore, but they have received very little rigorous analysis as tests indicative of the presence of underlying surgical disease.

In the terminology of evidence-based medicine, what the clinician who must decide about further investigations wishes to know is the positive predictive value (PPV) of these cutaneous signs. The PPV is the probability of disease in the patient in whom a test is positive, in this context, in the patient with a certain physical finding. Clinical experience suggests that the PPV of the classic lozenge-shaped lumbar hairy patch is very high. My impression is that nearly all such patients harbor underlying split cord malformations. The PPV of the flat, lumbosacral, capillary hemangioma ("salmon macule" or "strawberry nevus") is much more problematic.

The treacherous aspect of the concept of PPV is that it is not an intrinsic property of a test. Sensitivity and specificity are intrinsic properties, but, unless the specificity is perfect, the PPV of a test is dependent on the population of cases to which it is applied. If the prevalence of the target condition is high in the study population, the PPV will be high; if the prevalence is low enough, the false positives will swamp the true positives, and the PPV will be low. In discussions of lumbosacral capillary hemangiomas, readers must pay close attention to the derivations of the study groups.

In this issue Tubbs, et al., describe 21 patients with OSD in whom the only cutaneous sign was an isolated capillary hemangioma. These 21 cases were culled retrospectively from the pooled personal series of three pediatric neurosurgeons treating a total of 120 cases. Symptoms, neurological signs, and reasons for referral are not presented. Logically, one can conclude from this report that there exist patients with isolated capillary hemangiomas in whom OSD is present. Determination of the PPV of isolated capillary hemangiomas in a defined population was not the object of this study.

In contrast, we recently reported the yield of positive ultrasonographic findings in a small series of infants who were referred for pediatric neurosurgical consultation because of lumbosacral capillary hemangiomas (or strawberry nevi, as we termed the lesion in question). We attempted to estimate the PPV of the isolated strawberry nevus in a reasonably well defined, clinically relevant study population. Among 14 infants with isolated strawberry nevi, there were no positive spinal ultrasonographic examinations, and in an additional infant the magnetic resonance (MR) imaging study was negative. We estimated the PPV of the isolated strawberry nevus in terms of ultrasonographic yield to be 0 with a 95% confidence interval ranging to 0.2. We concluded that ultrasonographic screening of infants with isolated strawberry nevi is probably not worthwhile, and we asserted that the strength of the association between strawberry nevi and OSD remains unquantified.

Reconciling our observations with the observations of Tubbs, et al., is not difficult. Because the sensitivity of spinal ultrasonography is not perfect, there may have been infants with OSD in our study group who escaped detection. Neonatal strawberry nevi at other locations, such as at the glabella, on the eyelid, and in the subocciput, fade variably with time. Many of the lumbosacral strawberry nevi in our study group likely faded later in childhood as well. Perhaps inconsequential lesions tend to fade and significant lesions persist, in which case the PPV of an isolated nevus in an older population would be greater than in a younger population. We do not know.

Tubbs, et al., make an unqualified recommendation that lumbosacral hemangiomas be evaluated with MR imaging. This recommendation reflects expert opinion and common practice, but its evidentiary basis is very slim. The cost of such a recommendation is unknown but, because of the large number of infants with lumbosacral hemangiomas, probably quite high in terms of dollars and sedation-related misadventures. The benefit is unknown as well, in view of the undocumented natural history of...
most of the lesions mentioned in their report. The authors call for prospective studies of MR imaging in affected infants, as we did in the conclusion of our report, and let me now add as well a call for prospective, objective measurements of clinical outcomes in both treated and untreated asymptomatic infants and children with filum lipoma, meningocele manqué, terminal syrinx, and other minor forms of dysraphism.

Modern clinical research methodologies have much light to shed on the management of OSD.

References

RESPONSE: In reply to the editorial regarding our paper, we wholeheartedly agree that retrospective studies are intuitively flawed and that the most valid approach to this and every other question in a utopian world would be a randomized prospective trial. Our situation here, however, is not utopian. Neurosurgeons have recognized for more than 40 years that there appeared to be an association between isolated lumbosacral flat capillary hemangiomas (LSFCHs) and surgically significant intradural disease. Common practice in most centers is to evaluate these patients and offer surgical intervention when significant disease is thought to be present. We agree with Dr. Piatt that a focal area of hirsutism over the spine has a very strong correlation with split cord anomalies, and, furthermore, that of the cutaneous signatures of occult dysraphism with the least reliability is LSFCH. The disagreement rests in what we do until the definitive trial is performed.

Dr. Piatt has proposed that this patient population not be evaluated and that we simply wait for clinical change to occur before evaluation and intervention occur. This change in routine practice is recommended based on 15 negative studies in patients with LSFCH.

In response to this publication we reviewed data obtained in a large group of patients who underwent surgery for significant OSD and determined the number in which LSFCH was the only finding. This yielded 18% of our total group. Had we adopted a conservative approach, this group would have then been allowed to present with neurological deficits later in life.

Not included in this study was a significant group of patients seen during the same period with isolated LSFCHs in whom evaluations were negative for disease. Without quantifying this group, families are routinely told that the yield in the LSFCH group of patients is less than 10%. This is not too disparate from Piatt’s findings.

The reader is then left to choose between evaluating this group with a “relatively” low yield of surgically significant disease or allowing the disease to evolve naturally. We would be more willing to adopt a conservative approach if the reversibility of the presenting neurological deficits were high; however, neurogenic bladder symptoms in particular are not.

The cost of obtaining numerous MR images that turn out to be negative must then be weighed against the ongoing cost of a fixed neurological injury that frequently can be avoided with prospective intervention.

Without reliable data from a large group of patients, we are uneasy with changing this routine evaluation despite its relatively low yield and significant cost. Doing so will likely cause injury to a significant group of children.

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