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**Syringohydromyelia with Van Buchem disease.**

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# LETTERS

## Syringohydromyelia with Van Buchem Disease

We describe a case of syringohydromyelia in association with a rare bone dysplasia, Van Buchem disease. The sclerotic calvarial thickening characteristic of this disease may have caused a small posterior fossa with herniation of the cerebellar tonsils, leading to syringohydromyelia.

A 37-year-old woman with no relevant family history presented with dissociative anesthesia of the upper limbs, distal muscle weakness of the hands, and lower motor neuron cranial nerve deficits. She had frontal bossing, hypertelorism, and prognathism. The clinical findings suggested syringohydromyelia. Magnetic resonance (MR) imaging revealed a thick calvarium with loss of the medullary cavity, cervical canal stenosis, bilateral cerebellar tonsillar herniation, and syringohydromyelia from C-2 to the level of the conus (Fig 1). Subsequent skeletal survey showed diffuse uniform bone sclerosis with endosteal thickening, mainly affecting the calvarium and base of the skull, with less prominent involvement of the mandible, ribs, vertebrae, and pelvis. The appendicular skeleton was less affected, with mild periosteal nodularities. The tubular metacarpals, medially widened tubular clavicles, and thickened mandible with increased angle were the only modeling defects. The clinical presentation and skeletal findings pointed to a diagnosis of Van Buchem disease (1). At surgery, a syringosubarachnoid shunt was placed at the D11–12 region. Posterior fossa decompression was not considered in view of the difficulty of removing the sclerotic bone.

The causes of syringohydromyelia are well known (2). Many bone dysplasias such as achondroplasia and trichorhino-phalangeal syndrome type 1 are known to be associated with syringohydromyelia. Although the small posterior fossa might explain the development of the syringohydromyelia, the possibility of a primary Chiari I malformation as an incidental association cannot be entirely excluded.

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## Comment

There are 14 named sclerosing dysplasias of bone. As a group, they are generally poorly understood and in most, the etiology remains obscure. These disorders reflect abnormalities in the formation and modeling of bone. The developmental errors can occur at the sites of both endochondral and intramembranous ossification. Diagnosis can be complicated by the rare coexistence of two or more sclerosing dysplasias in one patient.

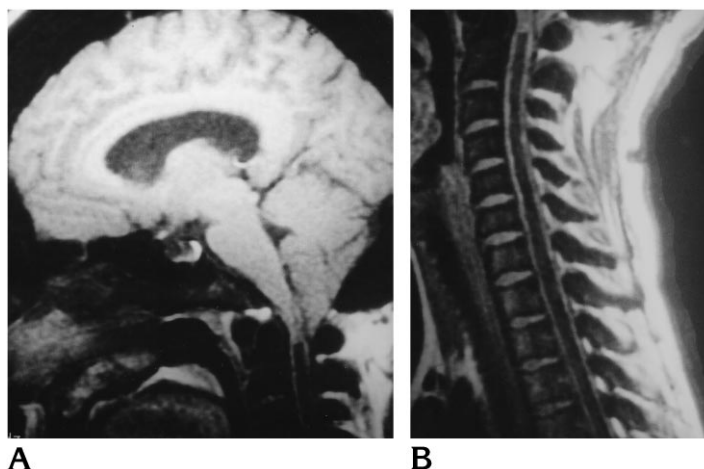
Van Buchem disease belongs to the endosteal hyperostosis subgroup of sclerosing dysplasias, and is characterized by defective intramembranous ossification. The clinical and radiographic differentiation between van Buchem disease (autosomal recessive) and its autosomal dominant counterpart, Worth-type endosteal hyperostosis, can be difficult. Both diseases show diffuse symmetric endosteal hyperostosis of the long and short tubular bones and mandible, and sclerosis of the skull, shoulder and pelvic girdles, and thoracic cage. Two features of van Buchem disease that help distinguish it from Worth disease include more severe mandibular involvement and the presence of small periosteal excrescences in affected long bones.

Neurologic complications in van Buchem disease include cranial nerve encroachment by hyperostotic bone (most commonly involving the facial canal, internal auditory meatus, and, less commonly, the auditory ossicles), cerebellar compression caused by reduction in size of the posterior fossa, chronic intracranial hypertension, and, rarely, spastic paraparesis. The genetic locus for the disorder has yet to be determined.

Dr Joseph's is the first report of syringohydromyelia in a patient with van Buchem disease. *Syringohydromyelia* refers to cystic dilatation of the spinal cord, and it may be congenital or acquired. The peak age at diagnosis is between 20 and 40 years. Specific symptoms and signs vary with the level of the lesion and its duration, because the disorder is progressive. Symptomatic syringohydromyelia is highly correlated with the presence of abnormalities of the posterior fossa, such as types I and II Arnold-Chiari malformations. Genetic syndromes with syringohydromyelia include myotonic dystrophy, Noonan syndrome, neurocutaneous melanosis, neurofibromatosis, Von Hippel-Lindau disease, and Fischer-Volavsek syndrome.

Syringohydromyelia also occurs with skull base deformity, and in association with progressive basilar invagination. The latter denotes an abnormally high vertebral column, with prolapse into the skull base. Basilar invagination is a feature of several genetic bone dysplasias including osteogenesis imperfecta, achondroplasia, and the rare Hajdu-Cheney syndrome. It has also been recognized

Fig 1. T1-weighted sagittal MR images of the brain (220/16/2 [repetition time/echo time/excitations]) (A) and cervical spine (300/16/4) (B) show thick calvarium, small posterior fossa, herniated cerebellar tonsils, and syringohydromyelia. The basal angle and foramen magnum dimensions are normal.



in Down syndrome when there is atlantooccipital instability.

The pathogenesis of syringohydromyelia remains poorly understood, but might be related to a disturbance in the dynamics of cerebrospinal fluid flow at the level of the posterior fossa. Thus, any disorder in which there is alteration of the normal anatomy at this site, whether by a tumor or by osteoporotic, osteolytic, or sclerotic bone, could potentially lead to development of a syrinx. In this context, it is not surprising that syringohydromyelia might occur in van Buchem disease. Rather, one might argue that perhaps it is not uncommon in the sclerosing dysplasias, but that only few are symptomatic.

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### Cerebellar Polymicrogyria

We read with interest "Brain MR in Fukuyama Congenital Muscular Dystrophy" by Aida et al (1). The authors described disorganized cerebellar folia with cystic changes as "cerebellar polymicrogyria." From our viewpoint, however, this name is not appropriately applied to such cerebellar lesions in Fukuyama congenital muscular dystrophy (FCMD), because there are not gyri but folia in the cerebellum. In addition, MR and pathologic findings of cerebellar polymicrogyria are not identical to those in polymicrogyria. Polymicrogyria is characterized by thick cortices with small convolutions and shallow sulci, and bumpy gray-white matter interface (2), whereas in cerebellar polymicrogyria a smooth cerebellar surface associated with small cystic lesions is seen (1). Microscopically, disorganized cortical laminar structures with or without an abnormal four-layered pattern are seen in polymicrogyria of the cerebrum (2). In cerebellar polymicrogyria, normal cortical layers (molecular layer and granular layer) are

maintained, although they consist of complex three-dimensional structures surrounding central pial tissue and vessels (3). Such histologic findings suggest that pathogenesis of the cerebellar polymicrogyria is different from that of the polymicrogyria of the cerebrum.

We would like to propose the term *polymicrophyllia* to denote the cerebellar lesion seen in FCMD (phyllon [φύλλον] is a Greek word representing the folium). This term may easily remind us of disorganized cerebellar folia, which are not identical to polymicrogyria.

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3. Takada K, Nakamura H. Cerebellar micropolygyria in Fukuyama congenital muscular dystrophy in fetal and pediatric cases. *Brain Dev* 1990;12:774-778

### Reply

We are grateful to Sasaki et al for their comments. As they suggest, the pathogenesis of polymicrogyria in the cerebellum may be different from that in the cerebrum. However, there must be causal multiplicities even in the

cerebral polymicrogyria (1). Furthermore, there is no evidence that the pathogenesis of cerebral and cerebellar polymicrogyria in FCMD is different. Sasaki et al postulate that cerebellar *polymicrophyllia* is a more appropriate term for the cerebellar abnormality in FCMD than cerebellar *polymicrogyria*. Their proposal may be relevant, signifying cerebellar malformation in FCMD from the semantic standpoint; nevertheless we do not prefer the term for two reasons. First, scientific terms should be comprehensible not only to specialists but also to nonspecialists; it would be difficult for the nonspecialist to understand the pathogenesis of cerebellar anomalies in FCMD from the Greek term suggested. Second, although even pathologists have noticed the nomenclature used for describing cerebellar cortical dysplasia to be inaccurate, the histologic findings of the cerebellum in FCMD have been called cerebellar *polymicrogyria* or *micropolygyria* in the pathologic literature of FCMD (2, 3, 4). We believe that semantics are important for the radiologist. However, the nomenclature used for describing cerebellar cortical dysplasia might be beyond their interest. Regardless of the semantics, the cerebellar malformation in FCMD represents the result of mutations of the FCMD gene, which would likewise create cerebral dysplasia. Further molecular investigation on FCMD, along with the genotype-phenotype correlation, may help elucidate not only the pathogenesis but also the clinical subdivision, which would be useful in patient care. Neuroradiologists should attempt to delineate in more detail the morphology of the complex central nervous system malformations that are mostly genetic diseases and wait for the molecular clarification.

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**Editor's note.**—The letter from Dr Sasaki et al was sent to A. James Barkovich for further review. His comments follow.

## Comment

Drs Sasaki, Ehara, and Watabe raise an interesting point. Although the folds in the cerebellar hemispheres and vermis are often referred to as *gyri* and *sulci* in common practice, the proper anatomic terms are, indeed, *folia* and *fissures*, respectively. In that sense, the authors' suggestion that the malformation seen in the cerebellar cortex of patients with FCMD be called *polymicrophyllia* is a reasonable one. However, their suggestion that the name should be altered because the MR and pathologic findings in the cerebellum of patients with FCMD are different from those seen in cerebral polymicrogyria raises a more complicated issue. As is pointed out in their reference 2, polymicrogyria is not a single entity, even in the cerebral cortex. Even in the broadest terms, cerebral polymicrogyria is divided into two major categories: layered and unlayered polymicrogyria. However, many other variations are seen pathologically. These include four-layered cortex with lamination, four-layered cortex without lamination, polymicrogyria with fusion of the molecular layers of the miniature gyri, and three-layered polymicrogyria (a molecular layer, a densely packed layer, and a sparsely packed layer beneath it) with poor lamination, in addition to other intermediate variations (1). In fact, the only feature that all cases of polymicrogyria have in common is multiple small gyri on the surface of the cortex when it is viewed under a microscope. Moreover, polymicrogyria is clearly etiologically heterogeneous, being seen in many genetic anomalies (including FCMD [2], Zellweger syndrome [3, 4], Neu Laxova syndrome [5], Aicardi syndrome [1, 6, 7], and Walker-Warburg syndrome [8, 9]), destructive lesions such as congenital infections (10) and in utero ischemia (11, 12), and miscellaneous anomalies with no known genetic associations (1). Finally, two different histologic variants of cerebral polymicrogyria have been described in patients with FCMD (2). It is clear, therefore, that *polymicrogyria* is a term used to describe a spectrum of malformations that are the end stage of a number of different processes. When this fact is considered together with the common usage of the terms *gyri* and *sulci* in referring to the cerebellar cortex, the use of the term *cerebellar polymicrogyria* to describe the irregularly interlaced islands of the molecular and granular layers seen in the cerebellar cortex of patients with FCMD no longer seems so unreasonable. Moreover, the term *cerebellar polymicrogyria* is commonly used and is in no way ambiguous. Perhaps Drs Sasaki, Ehara, and Watabe will agree that, although many of the terms that we commonly use in practice are not technically correct and they may offend those of us who are purists, it is sometimes easier to use them, secure in our knowledge that they are not perfect, than to try to change the habits of all of our colleagues.

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## Indications for Removal of Optic Nerve Sheath Meningioma

The radiologic-pathologic correlation by Ortiz et al (1) has beautiful radiographic images and histologic color pictures of an optic sheath meningioma. But the clinical practice of sacrificing useful vision (20/40) to get this specimen must be questioned. Why take out this tumor? The diagnosis is clear from the imaging studies. The tumor is benign. Later cure will not be adversely affected by watching the tumor and its images while the patient enjoys remaining vision. The proptosis is not reported in measurements but surely appears to be minimal and must not have been cosmetically serious. Diplopia was not present. The only selling point for surgery must have been the proposal for a cure otherwise thought to be unobtainable—a very doubtful proposition in my opinion. One hopes the patient was fully informed of the choices before surgery.

Because imaging is so important in neurologic, ophthalmic, and otologic diseases today, the *AJNR* is read by clinicians as well as radiologists. Clinical indications are more important than pretty pictures.

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## Reply

The choice to remove the tumor was made by the patient after being thoroughly informed of all of her options. Given the natural course and treatment options for this lesion, as mentioned in the article, our ophthalmologists strongly advised that the patient's tumor should be followed clinically and radiographically. Nevertheless, the patient insisted that the tumor be removed.

We are in agreement with the principles of informed consent and respect this patient's preference to undergo a surgical procedure (despite multiple attempts to convince the patient otherwise).

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**Editor's note.**—The letter from Dr Whittaker was also sent to Thomas Mizen. His comments follow.

## Comment

I agree with the comments of Dr Whittaker that the presentation by Ortiz et al represents an excellent clinico-pathologic correlation of the radiologic appearance and pathology of an optic nerve sheath meningioma. The recommended clinical management of a suspect meningioma is conservative with expected slow growth and progressive optic neuropathy. Clinical difficulties arise if the diagnosis is suspect, or if intracranial extension is identified; neuroradiologic imaging with contrast-enhanced MR is mandated for both the diagnosis and the monitoring of suspect optic nerve sheath meningiomas (1). A surgical approach to biopsy would be indicated for an orbital mass lesion with a more rapid progression than expected for meningioma, and resection would be indicated for a more malignant lesion identified at biopsy, which might extend intracranially along the course of the optic nerve. I would

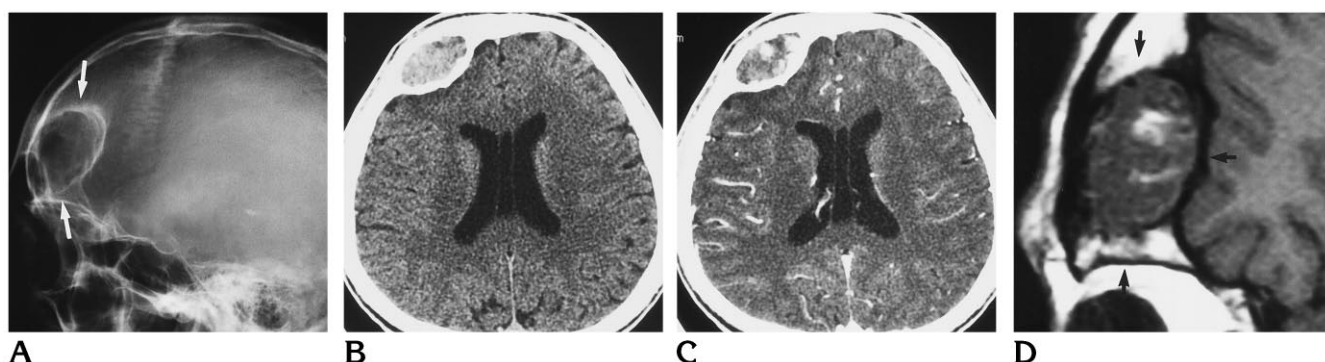


Fig 2. A 32-year-old man with history of trauma to the right frontal bone, intermittent headaches, and a right frontal bone mass. A, Plain radiograph of the skull (lateral) shows an osteolytic lesion with a well-demarcated sclerotic margin in the right frontal bone (arrows). B, Axial CT shows high density within the mass and C, patchy enhancement after contrast administration. D, Sagittal T1-weighted MR image (291/13) shows a spherical intradiploic mass with heterogeneous signal intensity (arrowheads).

agree with Ortiz et al that the natural course of optic nerve sheath meningiomas is slow progression. However, in my opinion, there is not considerable controversy in the management of optic nerve sheath meningiomas; since surgical excision cannot preserve vision, the decision for surgical excision would depend on the clinical exam (progressive optic neuropathy and proptosis) and neuro-radiologic imaging for intracranial extension (2). The sacrifice of vision should occur as a result of the natural progression of the tumor rather than the knife of the surgeon.

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## Intradiploic Hematoma

A 32-year-old man was referred for evaluation of a mass lesion in the right frontal bone. He reported intermittent headaches at the right frontal region, and had a history of minor head trauma at the same site when he was 13 years old.

Physical and neurologic examinations were normal. Plain radiographs of the skull showed a  $6 \times 4 \times 3.5$ -cm osteolytic lesion with a sclerotic margin (Fig 2A). Com-

puted tomographic (CT) scans showed that this lesion expanded inward and had a central high-density area with patchy enhancement after administration of contrast material (Fig 2B, C). MR imaging revealed a spherical intradiploic mass with heterogeneous intensity on T1-weighted images (Fig 2D). There was no invasion of underlying brain tissue. The intradiploic mass was resected. The pathologic diagnosis was organized hematoma of the skull.

Intradiploic mass lesions are uncommon. Yuasa et al (1) described an intraosseous hematoma of the skull in the left parietal region in a 20-year-old man with a history of remote head trauma, and Sato et al (2) reported chronic diploic hematoma of the parietal bone in a 20-year-old man who also had a history of head trauma. Our patient was also a young man with a history of minor head trauma. Although these authors postulated that the slowly progressive enlargement of this lesion was caused by organization of hematoma, its etiology remains controversial (1-3).

Similar pathologic lesions can be found in aneurysmal bone cyst (ABC), giant cell reparative granuloma (GCRG), and traumatic bone cyst (TBC), and the radiographic findings are also similar. Lichtenstein (4) has described the ABC as a primary lesion arising from traumatic anomalous venous disruption in the diploë with a subsequent increase in venous pressure and development of a dilated and engorged vascular bed in the affected bone. The clinical presentation is characterized by rapid enlargement of the mass under hemodynamic pressure (5). Jaffe et al (6, 7) have speculated that GCRG results from a local reparative reactive process related to a traumatic intraosseous hemorrhage or a periosteal reaction. However, the mechanism of enlargement in GCRG is still obscure (6, 7). Its clinical course is more gradual than that of ABC. TBC has been attributed to intraosseous hemorrhage after mild trauma (6). The cause of expansion of TBC has been suggested to be transudation into the encapsulated intraosseous hemorrhage resulting in pressure erosion of the bone (6). Al-

though the clinical entity of intradiploic hematoma remains controversial, it is close, if not identical, to TBC from both clinical and pathologic observations.

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## Hemisphere Activation during Motor Tasks

Although able in many cases to indicate the location and size of intracranial lesions, the radiologic procedures usually used clinically with acute stroke patients provide rehabilitation professionals with only limited insight into how the brain works and how far patients will recover functionally (1, 2). The imaging information provided is typically less instructive to clinicians involved in rehabilitation than that provided by simple clinical observations and measures of impairment and disability (3-5). Procedures such as functional MR imaging, on the other hand, have great potential to explain both the functioning of the brain and the motor performance originating from it. The purpose of this letter is to elaborate and comment on the observations made by Li et al (6), who used MR imaging.

Li et al noted brain activity that was both ipsilateral and contralateral to motor tasks involving each of their subjects' hands. They suggested that such "bilateral activation may explain ipsilateral and contralateral impairment in patients with unilateral hemispheric lesions." Li et al cited several references supporting the bilaterality of activation. Further support for their suggestion can be found in a further analysis of their data. The numbers of activated pixels they reported to be generated within the two hemispheres are highly correlated during both left and right hand motor tasks. Using their data, I calculated Pearson's correlations for ipsilateral and contralateral activation to be .829 for motor tasks involving the left hand and .887 for motor tasks involving the right hand. Additional

support for their suggestion is provided by recent research on muscle strength after stroke (7). That research documented weakness ipsilateral to stroke that was greater proximally than distally. Such a finding is consistent with the proposal of Gandevia that ipsilateral weakness should be greater proximally (8). Relevant also is recent research showing that strength measures are correlated significantly between sides ( $r = .540$  to  $.833$ ) (7). Muscle strength measures also have a high degree of internal consistency across actions and sides, whether obtained from patients with stroke or healthy persons (9). Taken together, the findings of Li et al and those of others suggest a parallel organization of action based on the overall goal of the motor act (10).

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## Reply

Dr Bohannon, a physical therapist, raises two interesting and important points regarding the data that we reported.

Functional imaging shows that normal human motor function requires parallel distributed networks involving separated but functionally related regions of the brain. Bressler (1) emphasizes that M1 is not a final integration center for movement, but that multiple cortical areas work together as a distributed network. With functional MR imaging, Sanes et al (2) showed that a large expanse in the human precentral gyrus (M1) exhibits activation during individual finger or wrist movements and that the regions of activation for finger or hand overlap. These studies are interpreted as showing a distributed and cooperative network simultaneously controlling collections of muscles (2). The distribution of motor function between the two hemispheres probably varies among individuals. Some evidence suggests that bilateral representation of motor function diminishes with age. Contribution of the ipsilateral hemisphere to motor function may explain the occasional patient who has an ipsilateral paresis during the Wada test. It may explain recovery of motor function after cerebral infarction.

Secondly, Bohannon raises the question that functional MR may be used to study mechanisms by which patients recover function after cerebral infarction. A number of mechanisms have been proposed to account for recovery: redundancy in neural representation of function, sprouting and reinforcement of existing though normally secondary neuronal circuits, and persistence into adulthood of bilateral cortical control of motor function. A positron emission tomographic study in patients recovering from stroke showed that when the recovered hand was moved, cerebral activation was bilateral to a greater degree than in healthy subjects. The study suggested a role of ipsilateral cortical efferent pathways subserving movement after contralateral injury (3).

We thank Dr Bohannon for his letter. With the interest and cooperation of clinical colleagues, investigators may be able to develop functional MR as a means to determine the prognosis from stroke and to select patients for rehabilitation therapies.

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**Editor's note.**—Additional review of the paper by Li et al was sought from Dr Lewine and colleagues. Their comments follow.

## Comment

Several studies of brain hemodynamics using positron emission tomography and functional MR converge to indicate bilateral activation of sensorimotor cortex during the performance of unilateral hand movements, especially when the nondominant hand is involved (1-2) (Mattay VS, Santha AKS, Van Horn JD, Sexton R, Frank JA, Weinberger DR, "Motor Function and Hemispheric Asymmetry: A Whole Brain Echo Planar fMRI Study," [abstract] and Falk A, Durwen HF, Mirzaian E, Müller, Gehlen W, Heuser L, "Evaluation of Bilateral Cortical Activation in fMRI with Unilateral Repetitive Hand Movements" [abstract], both in *NeuroImage*, Proceedings of Second International Conference on Functional Mapping of the Human Brain, Boston, Mass: 1996;3:S398). The recent study of Li et al replicates these findings, and it extends them by showing ipsilateral activation during a somatosensory task.

As the authors indicate, the motor findings are supported by several clinical reports of ipsilateral motor compromise from unilateral lesions (3-5). One of the most telling reports is that of Alf Brodal (6), the noted neuroanatomist, who had a right hemispheric stroke. In addition to a left-sided hemiparesis, Brodal noted the appearance of defective motor control of his right hand. Beyond clinical observations, basic science data also support the notion of bilateral motor cortex activation during unilateral movements. For example, extracellular recordings from the monkey primary motor cortex demonstrate about 8% of neurons to show activity before an ipsilateral digit movement (7).

The situation with respect to ipsilateral somatosensory activation is somewhat less clear, and we agree with Li et al that "these data must be interpreted conservatively because functional MR imaging techniques are evolving." To be sure, there are ipsilateral somatosensory projections and clinical evidence for ipsilateral changes in tactile perception after unilateral lesions (8, 9), but electrophysiological studies suggest that it is very difficult to drive ipsilateral primary somatosensory cortex directly using tactile stimuli. On the other hand, it is known that contralateral somatosensory cortex sends transcallosal projections to ipsilateral SII (secondary somatosensory cortex). Also, there are projections to the contralateral posterior parietal cortex, which in turn send transcallosal projections to the ipsilateral posterior parietal cortex. Given this complicated cascade for the processing of tactile information, care must be taken to clarify exactly what ipsilateral brain regions are activated by tactile stimulation. For example, in Figure 1 of Li et al, somatosensory stimulation gives activation of several areas in each hemisphere; one must also be careful to distinguish between the precentral gyrus, postcentral gyrus, posterior parietal cortex, and area SII, as all of these may be activated by somatosensory stimulation. Ipsilateral activation of primary somatosensory cortex clearly has different ramifications than ipsilateral activation of SII or posterior parietal cortex. Also, it is important to clarify the temporal dynamics of activation, which has additional ramifications for the interpretation of



the data. Direct ipsilateral activation has different implications from transcallosally propagated activation.

Although we emphasize these later points with respect to the somatosensory situation, they may be equally applicable to the motor data. For example, using magnetoencephalography, we have observed that some subjects performing unilateral digit movements show bilateral activation of primary somatosensory cortex (Davis JT, Lewine JD, Thoma R, et al, "The Post-movement Evoked Field: A Manifestation of Somatosensory Feed-back or Motor Feed-forward," Society for Neuroscience 1996;22:890 [abstract]). The timing of this activation suggests a bilateral "feedforward" of information from one motor cortex to both right and left somatosensory systems. The data are consistent with the concept of "efference copy." Given this situation, it is important to know whether the functional MR observation of bilateral activation is indicative of bilateral sensorimotor control of unilateral movements, or merely a reflection of one hemisphere informing the other about an action it is taking. Perhaps, through combined magnetoencephalographic/functional MR experiments, the spatio-temporal dynamics of ipsilateral and contralateral sensorimotor integration can be further elucidated and clarified.

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