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FROM THE ARCHIVES

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As a young neurologist, (Sir) William Gowers (1845–1915), working in collaboration with John Hughlings Jackson (1835–1911), described the case of Eliza Joad. First designated as 'imperception', the term 'agnosia' was subsequently introduced to describe her inability to perceive or interpret items that nonetheless were sensed. With the naming of 'apraxia' by Hugo Liepmann (1863–1925), 'neurologists . . . began to unravel the complicated tangle of frontal, temporal, parietal and occipital dysfunction'.

Macdonald Critchley introduces his William Gowers lecture with a quote from Andre Maurois: 'The members of the medical fraternity can at least classify our ailments in carefully labelled compartments, and that, in itself, is reassuring. To be able to call a demon by its name is half-way to getting rid of him'.

Noting Gowers' dislike of descriptive papers, Critchley opens his lecture with an account of the patient described by Josef Gerstmann (1887-1969), writing from Vienna in 1924 (*Wiener Klinische Wochenschrift* 1924: 37; 1010–1012), who—amongst other deficits—is unable to name her fingers and identify her own or the examiner's individual digits on request. He calls this symptom 'finger-agnosia' which Critchley considers to be 'an ill-chosen expression'. In 1927, Gerstmann describes two new cases and proposes that difficulty with writing forms part of the emerging syndrome. Further observations add yet more phenomena and, by 1930, the descriptive tetrad is complete: 'Gerstmann's syndrome' now consists of finger-agnosia, dysgraphia, dyscalculia and right–left disorientation; and it localizes to the left angular gyrus—a region assumed by Gowers to be a higher visual centre. A quote from Oscar Wilde, aimed at those interested in cerebral localization, who find 'a curious pleasure in tracing the thoughts and passions of men to some pearly cell in the brain, or some white nerve in the body, delighting in the absolute dependence of the spirit on certain physical conditions, morbid or healthy, normal or diseased' seems apposite. But this strange collection of symptoms also appeals to the more 'dynamically minded'. Inevitably, others have already described the individual features of Gerstmann's syndrome, but no previous author has gathered the four components together and recognized their autonomy, save one.

Jules Badal (1840–1929; see Fig. 1), the first person to hold a professorship of ophthalmology in France (Bordeaux) and inventor of an ingenious optometer and a refracting ophthalmoscope, has described the case of Valérie, aged 31 years, in whom the complications of eclampsia leave her unable to dress or feed herself, to read or write or manage eye–hand coordination, and with an altitudinal field defect. She can neither trace nor copy letters of the alphabet; she is muddled with respect to left and right; she cannot localize sounds; and 'if . . . asked to name the five fingers of the hand in their correct order, she rarely [does] so without some mistake, either in their names or in their classification from 1 to 5'. Dr Critchley knows of Badal's paper, ignored by previous commentators other than Arnold Pick (1851–1924), through the historical acumen of Dr Arthur Benton (1909–2006). But with rich speculation on the concept of Gerstmann's syndrome and its causes—focal lesions and congenital or constitutional deficiencies synonymous with developmental dyslexia—'Gerstmann's syndrome has recently become the object of increasingly damaging assault . . . culminating in a recent *expertise* which

Figure 1 Jules Badal (1840–1929).

[Benton] entitle[s] ''the fiction of Gerstmann's syndrome'' . . . and [the contribution] . . . from two neurologists from Freiburg, whose paper *Gibt es das Gerstmann-syndrom?* (''Is there such a thing as Gerstmann's syndrome?'') was delivered at a symposium in Baden-Baden'.

Now Dr Critchley considers each component of the tetrad in turn. The difficulty with finger naming is simply a defect of language (there being no sense of 'finger-gnosis' that can be lost in isolation) that, by 1957, Gerstmann extends to include the inability to recognize, identify, differentiate, name, select, indicate or orient the individual fingers of either hand belonging to the patient or someone else; and in the context of preserved intellect, speech, vision, perception, spatial orientation and motor and sensory function. Given the ever more complicated test procedures applied to these patients and the variations in their performances, terminology has become more fanciful: finger aphasia, optic finger-aphasia, constructive finger apraxia, apraxic disturbance in finger-selection, finger-sense, finger order-sense, finger-naming, finger-manipulation, finger-apraxia and finger-side localization. In general, the thumb and little finger are easier to identify than the middle three digits. Patients are examined for their finger-naming ability without cues from vision or speech, and using finger-like structures detached from the patient, such as models of hands and gloves. Interlacing fingers from the patient and another person (the Japanese illusion) adds yet more complexity; and even toe-agnosia must be distinguished. Dyslexic children are poor at finger-naming, although Macdonald Critchley considers that the expected age of acquisition of this knowledge (7.5 years) errs on the young side. Furthermore, there are national confusions in finger naming (English people owning a thumb and four fingers whereas Continentals have five fingers). Again, Gowers can be quoted (but from another context): '. . . a large number of new terms have been introduced, most of which are needless, and to some extent injurious, fostering a harmful tendency to divide where it is desirable only to distinguish'.

It has taken time for the association with right–left disorientation to be recognized as part of the putative syndrome. Here too, the unfortunate patient with Gerstmann's syndrome is tested for accuracy on complex and irrational sequential tasks ('touch your right ear with your left hand'). He or she is required to name the right and left sides of someone else seen in the mirror; or to orientate correctly parts of a doll held in all sorts of awkward postures. These are skills that may not develop normally until adolescence. However, in general, patients do better at lateralizing parts of their own body than other people's or objects in space. They may use cues, as did Badal's patient who liked to orientate herself by making the sign of the cross; and a taxi driver marked one thumb with a black cross in order to remember right from left and therefore to work out which route to take.

The defect in calculation has been relatively under-studied. It affects mental arithmetic and the completion of sums on paper; it affects ordering according to magnitude, counting backwards and sorting odd from even numbers. Macdonald Critchley considers spatial and dysphasic categories of dyscalculia and suggests that some subjects should be described as having 'anarithmia'—loss of the principles of mathematics. Dysgraphia, the fourth component of the 'quadrivium', involves both dyspractic and dysphasic elements. Penmanship is defective with poorly constructed letters and words, unattractive formatting on the page, and 'acopia' (from the ability to copy not to cope). Words are omitted or reproduced as perseverations—misspelled, invented and subject both to semantic and syntactical errors. Is there a unitary underlying defect in this syndrome?

Gerstmann considered finger-agnosia, one element of a more fundamental autotopognosia, to be the principal item; but why this should be confined to the hand is less easy to comprehend. Does the explanation lie in demotion of the hand as a highly evolved organ of touch and tool use, having additional paralinguistic attributes (as in gesture), to a more primitive role as a prehensile or feeding instrument analogous to the elephant's trunk or the monkey's tail? Gerstmann argues that 'the body schema [is] affected in . . . functionally the most significant, differentiated and vulnerable . . . sphere . . . concerned with the individual fingers – as though the optic-tactile-kinaesthetic image pertaining to the fingers were split off from the total body image, the finger scheme [detached] from the total body scheme'. Others put this is comparable terms: 'a deficiency in the finger schema' . . . 'the fingers fused into a single lump'. Alternatively, Gerstmann's syndrome is considered to be a variant of dyspraxia showing loss of digital dexterity and thereby also accounting for the dysgraphia.

However, Oliver Zangwill (1913–87) considers attempts to settle whether 'finger-agnosia ''causes'' finger-apraxia, or vice versa, [as] fruitless, since both types of behavioural impairment can be conceived as representing a single basic neuropsychological deficit'. There has followed the concept of Gerstmann's syndrome as a disorder in spatial thought in which the hand, disconnected from its directionality, no longer acts as a tool connecting personal and extra-personal space. That formulation sits uncomfortably with the preserved orientation of affected individuals with respect to vertical space, and concepts such as 'in' and 'out', 'far' and 'near', and 'above' and 'below'. Self-evidently, calculation and finger-play are connected. The Romans used 'digitum' to denote a number. Primitive peoples used the word 'five' to denote the 'hand'; 'six' was a 'hand plus one'; and 'ten' was 'half a man'. The Aristotelian 'decimal' system is based on 10 fingers; and the 'vingesimal' phrases of the Danish, Irish and the Basque tongues merely relate to 10 fingers and 10 toes. But, drawing on the concepts of (Sir) Henry Head (1861–1940), Benton has argued in favour of a basic defect in symbolic understanding, operation or expression that forms part of more generalized defect of propositional awareness—a formulation that Dr Critchley now aligns with his own concept of latent or 'pre-aphasia'.

Doubts began to emerge on the validity of Gerstmann's syndrome as a genuine entity soon after the original series of papers appeared. One or more of the individual components might be absent in the individual case, or appear in isolation. Furthermore, insufficient attention has been paid to features that commonly occur in association with the core symptoms—hemianopia, constructional and ideomotor apraxia, pain asymboly, achromoatopsia, dysphasia, generalized intellectual loss and abnormalities of timing. Perhaps, statistical analysis of how often the four cardinal symptoms appear together may be revealing. As for the core manifestations representing an expression of disease that exclusively affects the left angular gyrus,

Dr Critchley emphasizes that the larger the parietal lesion, the more likely the syndrome is to occur; although it may be seen in diffuse brain disease and in the context of lesions outside the parietal lobe. Dr Benton has studied cases over a period of >15 years and reluctantly concludes that the so-called Gerstmann syndrome is an artefact of the tests applied and nothing more than an aggregation of physical signs produced by any lesion large enough to involve contiguous brain regions. Others share his scepticism. Dr Heimburger and colleagues have studied over 100 cases and find that about a quarter of this group each manifests one, two, three or all four of the required features—the number increasing directly with the size of the underlying lesion and the probability of left lateralization: 'the syndrome does not appear as an isolated cluster of deficits against an otherwise normal neurological status. The associated deficits are an integral part of the total picture of nervous disability . . . a destructive lesion of the left angular gyrus is not a necessary condition for Gerstmann's syndrome'; and the area of tissue damage often extends into the supra-marginal gyrus as far back as the inferior parietal lobule, the superior temporal gyrus and the anterior part of the occipital lobe. In rank order, the frequency of the individual components is dyscalculia, then dysgraphia, right–left disorientation and, least commonly, finger-agnosia. That said, the two least common symptoms tend to occur together most often, whereas other combinations are apparently more random. More recently, Klaus Poeck (1926–2006) and Bernt Orgass have reported that, as the number of features increases, so too does the probability of additional cognitive deficits—especially constructional apraxia—also being present.

Against this background of an ambiguous syndrome of behavioural neurology, Dr Critchley asks himself about the status of the Gerstmann symptoms in developmental dyslexia. The relationship seems no less secure and, despite having himself promulgated this concept in the 1940s, Dr Critchley is now not at all persuaded. In developmental dyslexia: 'right-left confusion is but transient . . . the defect of writing is not a dysgraphia . . . dyscalculia is rare . . . and ''finger-agnosia'' is even more suspect than it is in the adult . . . as Wilde said ''it has all the vitality of error, and all the tediousness of an old friend'' '. Do other clusters of symptoms deserve to be clustered into a syndrome alongside those described by Drs Badal and Gerstmann? Should we acknowledge Leonard's syndrome—disorders of the sense of time, constructional apraxia, dysgraphia and dyscalculia; Zeh's syndrome—acalculia, dysgraphia and loss of recognition of facial expression in pictures; Cogan's syndrome—hemianopia, loss of optokinetic nystagmus, dyslexia, spasm of conjugate gaze as well as the four elements of so-called Gerstmann's syndrome; von Angylal's syndrome—right–left confusion, dysgraphia and thumb–mouth agnosia; and Stengel's syndrome—constructional apraxia, loss of spatial orientation and the four features of Gerstmann's syndrome? Pausing only to acknowledge Gerstmann's luck in writing at a time when neurology was sympathetic to the assignment of eponymous syndromes, Macdonald Critchley ends by again quoting Sir William Gowers: 'there are very few observations in medicine regarding which it is not obvious that they would speedily have been made by someone other than the actual observer; that it is very much of an accident that they were made by certain individuals. Scientific nomenclature should be itself scientific, not founded upon accidents. However anxious we may be to honour individuals, we have no right to do so at the expense of the convenience of all future generations of learners'.

Does modern clinical neuroscience also distrust the entity of Gerstmann's syndrome as a discrete cluster of symptoms having a common underlying neuropsychological basis and localizing to the left angular gyrus? On page 320, Elena Rusconi and colleagues review the concept and conclude that whilst 'it is legitimate to label the conjunction of symptoms first described by Gerstmann as a ''syndrome'', this cluster does not result from damage to contiguous neurons subserving the properties that are disrupted but, rather, from disconnection of co-localized fibre tracts in the subcortical parietal white matter'.

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