Dear Editor

Everyone knows that autism is a primary brain disorder. This top-down theory seems so self-evident that it has proved impossible to get autismologists to engage with let, alone test, a radically different bottom-up theory for which there is a large amount of evidence, namely that autism is a variant ear disorder (Gordon 1989). This means that every cause of autism is also a cause of deafness, any brain malfunction being secondary to sensory misspecification. Two recent papers provide a powerful comparison of these rival theories.

Mankoski et al., (2006) provide clear evidence for autism being caused or aggravated by malaria, but totally ignore the fact that this was predicted by the otogenic theory, as is the case with their other implicated causes of autism (herpes, rubella, meningitis, mucopolysaccharidoses and cranial dysostoses) which also cause deafness (Gordon, 1989). One of their cases became autistic after Salmonella meningitis. Deafness is prominent after meningitis in African infants: of 19 survivors, 5 had sequelae, 3 of whom were deaf (Molyneux et al., 2000). Not all causes of deafness may also cause autism, but there is a very long list of them that do (Gordon, 2007). So, does it include malaria, and is not the autism more plausibly explained by associated brain damage?

Malaria is a common cause of deafness in Africa (Olu Ibekwe, 1998). Despite the strong co-morbidity between deafness and autism, most autists have normal pure tone audiograms (Hayes and Gordon, 1977). Hence any causal relationship is not with hypoacusis, but must be with hyperacusis or auditory fluctuation or distortion, as evident clinically and from many written accounts by autists. Itard (1821), who coined the word hyper(a)cousie, noted that this was often the first sign of progressive cochlear hypoacusis. There is a clear otological candidate for such an auditory syndrome, Meniere Spectrum Disorder, comprising the usual symptoms of Meniere’s disease, but not chronic, obvious or progressive enough for a diagnosis of Meniere’s disease in an otology clinic. The immediate trigger for this inner ear hyperirritability seems to be a drop in perilymph pressure from a fistula, low blood or spinal fluid pressure, weight loss, etc. (Gordon, 1983). In malaria, dehydration would be a clear trigger of such an endolymphatic hydrops. A doctor in Africa (Denti di Pirajno, 1956) gave an excellent description of his own sudden malarial Menieriform attacks: his knees turned to water; slight buzzing in his ears grew into deafening roar; he experienced appalling nausea; he seemed to fall through a whirl of sparks into blackness and unconsciousness.

As for brain lesions causing autism,
only 1 of their 7 cases of malarial autism had cerebral malaria, far fewer than expected if brain damage was crucial. Case G had mild malaria without sudden effect, ruling out an acute brain lesion. A more plausible developmental scenario was malaria attacking a genetically vulnerable cochlea (his father was deaf), causing gross auditory distortion, his gradual speech loss and autism (erroneous auditory feedback being more disruptive than none). Malaria causes language disorders, including autistic semantic pragmatic impairment. Carter et al., (2006) found an odds ratio of 3.7 for speech/language disorder after cerebral malaria, but also odds of 3.1 for non-cerebral malaria, confirming their previous “striking observation” of no difference in language ability between those with cerebral and non-cerebral malaria. The evidence against an encephalogenic cause of autistic communication disorder does not prove an otogenic cause, but is consistent with it.

A further comparison comes from two autistic males with Waardenburg syndrome (Kiani et al., 2007). Both were profoundly deaf, like only 1 of 22 with Waardenburg syndrome from a specialist neurotology clinic (Black et al., 2001), confirming autism-deafness comorbidity. Other pigmentary disorders with autism and deafness include retinitis pigmentosa (Gordon, 1991) and albinism (Mankoski et al., 2006). As noted above, the hypoacusis per se is more a clue for, than cause of, autism. In fact, only 4 of the 22 had a chief complaint of hearing loss. On the other hand, they had a full house of fluctuant symptoms of hydrops; dizziness (90% of cases), tinnitus (85%), disequilibrium (75%), vertigo (75%), aural fullness/pressure (75%), headache (75%), nausea (65%), visual sensitivity (65%), noise intolerance (65%) and subjective hearing loss (40%). Of the 15 with normal pure tone audiograms, all had objective vestibular abnormalities, 80% had abnormal electrocochleography, and at least 13% had subjective hearing loss. None were diagnosed as having Meniere’s disease. Signs of Waardenburg’s syndrome may be quite subtle, so it is easily missed in otology clinics, even more likely so in autism clinics (although Mankoski et al., 2006 noted some hypopigmentation in Case D). Kiani et al., (2007) found limited evidence for mental retardation in Waardenburg’s syndrome, and reported no evidence of brain damage in their 2 cases. Black et al., (2001) note that mental retardation is rarely reported, most often in association with deaf-mutism and failure to learn and interact, raising the possibility of confusion between sensory and intellectual disability.

Meniere spectrum disorder is clearly recognisable in many autobiographies (e.g. Williams, 1999). “I began to get dizzy constantly…I began to lose weight”. “I knew this monster. It was the Big Black Nothingness…My ears hurt…My knees went to the floor…My head seemed to explode…Dizziness and exhaustion began to overtake terror…You must escape because you hear the sound of ‘tidal waves’”. “My emotions overwhelmed me, my hearing became painfully acute, and the meaning fell out of everything…The muscles in my ears contracted with the amplified sound of rushing blood to add to the meaningless cacophony”. “It gave me a headache when my senses got flooded”. “My head was swimming and I felt nauseous”. “I was surrounded by colour and pattern and shape, with my sense of hearing heightened, my sensitivity to light increased”. “Jim and I both had trouble with sensory overload and shutdown through our eyes and ears”. “I heard noise but was deaf to most of the meaning”. “Autistic people existed whose primary difficulties were sensory ones. There were those who had trouble standing the world as well as understanding it”.

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She reports all ten diverse hydrops symptoms as listed by expert otologists (Black et al., 2001).

Many genes for autism must also be genes for deafness, and in both conditions the genes are often unlocked by exogenous factors. In Africa (Olu Ibekwe, 1998) genetic deafness, like autism, often presents after non-cerebral infectious illness.

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References


