Review article

Mutism: elective or selective, and acquired

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Abstract

When a child does not speak, this may be because there is no wish to do so (elective or selective mutism), or the result of lesions in the brain, particularly in the posterior fossa. The characteristics of the former children are described, especially their shyness; and it is emphasized that mild forms are quite common and a definitive diagnosis should only be made if the condition is significantly affecting the child and family. In the case of mutism due to organic causes, the commonest of these is trauma to the cerebellum. Operations on the cerebellum to remove tumours can be followed by mutism, often after an interval of a few days, and it may last for several months or longer, to be followed by dysarthria. Other rarer causes are discussed, and also the differential diagnosis. The so-called posterior fossa syndrome consists of mutism combined with ataxia, cranial nerve palsies, bulbar palsies, hemiparesis, cognitive impairment and emotional lability, but the post-operative symptoms are often dominated by the lack of speech. The most accepted cause for the condition is vascular spasm with involvement of the dentate nucleus and the dentatorubrothalamic tracts to the brain-stem, and subsequently to the cortex. Diaschisis may be involved in causing the loss of higher cerebral functions, and possibly, complicating hydrocephalus. The treatment of elective mutism is reviewed, either using a psychotherapeutic approach or a variety of drugs, or both. These may well be ineffective, and it must be remembered that the condition often resolves on its own. The former treatment must concentrate on the training of social skills and activities of daily life and must be targeted to both the child, the family, and the school. Also, all kinds of punishment and insistence on speech must be discouraged. The drug, which seems to be most effective, is fluoxetine. Discovering more about the causes of mutism due to organic causes may well depend on studies using such techniques as magnetic resonance imaging and single photon emission tomography. © 2001 Elsevier Science B.V. All rights reserved.

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1. Introduction

When a child does not speak there are two possible causes. The child may choose not to, in which case it is elective or selective mutism, similar terms for the same condition or organic causes may prevent speech, as in the posterior fossa syndrome. In both instances, it is likely to cause anxiety to those caring for the child.

2. Elective or selective mutism

Many children in their early years do not speak more than a few words to those outside their immediate circle of friends and relations, so that elective mutism can be considered as one end of a naturally occurring spectrum. For example, such children may talk a great deal at home, but little, if at all, at school. In a series of children with this condition, it was shown by Steinhausen and Juzi [1], working in Zürich and Berlin, that it typically starts at pre-school age, is more common among girls than boys, and is seen in all levels of society. Early developmental risk factors are quite common, and so is a history of immigration; and unfamiliar people and the school environment were the most frequent context in which the children do not speak. In up to one third of the patients, there was a history of premorbid speech and language disorders, especially those of expressive language and articulation, and three-quarters of them had behaviour disorders in infancy and early childhood. As might be expected, the children were often shy and anxious, and sometimes withdrawn.

In a screening of 7–15-year-olds over a full school year for selective mutism in Sweden, Kopp and Gillberg [2] found an incidence of 18 in 10 000 children, which was higher than in some previous reports. This may be due to the fact that some parents of these children are themselves shy and reluctant to report their child’s condition. Also, there were 89 in 10 000 children who were shy and reticent, but did not fulfil the diagnostic criteria for mutism. There were more girls than boys in the later group, but not in the
former, although the numbers were too small to draw definite conclusions on this, and on whether the two groups represented separate entities.

The disorder has been reported among twins, but it is not known for certain if it occurs more frequently; although it may well be reported more often because of its rarity [3]. It has also been noted in two sisters with the fragile X syndrome, both of whom were anxious and shy [4]. Although there may be no direct link between the mutism and the abnormality in the FMR1 gene, selective mutism may be more common in children with this syndrome than in the general population because of its clinical features.

3. Organic causes of mutism

If a child is mute, or says very little in any environment, the possibility of a lesion of some kind in the speech pathways has to be considered. The history and examination may well indicated that there is such cause, but sometimes this can only be established when psychogenic causes have been excluded and relevant investigations have been done.

It is now recognized that lesions of the cerebellum can cause speech disorders [5]. In particular, disorders such as mutism can be a complication of posterior fossa surgery, but rarely occur after other cerebellar injuries, such as cerebellar haemorrhages, subarachnoid haemorrhage [6] vertebral artery injuries, basilar artery occlusion [7], and head trauma. In a series studied by Janssen et al. [8] of 21 children who had undergone surgery for large posterior fossa tumours during a period of 4 years, six developed post-operative mutism. There is typically a period of normal speech immediately after surgery, and many of the reported patients had no previous problems with language. There were often associated problems of dysphasia and emotional lability. The speech disturbance lasted from a few days to several weeks, and recovery might take from a few days to several months, but most returned to normal speech. Some of the affected children were described as being hoarse and hypernasal or with a strained and strangled vocal quality after the mutism, and dysarthria is common. This raises the possibility that in some cases, the post-operative mutism may be an extreme form of cerebellar dysarthria [9]. There may also be high-level linguistic deficits after surgery, such as in processing speed, memory, and cognitive planning. It is accepted that the cerebellum co-ordinates the articulatory muscles, and any impairment of function will lead to dysarthria, but recent research has definitely shown that it will contribute to language and cognition [10]. In a study on 20 patients with diseases confined to the cerebellum by Schmahmann and Sherman [11,12], it was found that if the posterior lobe of the cerebellum and the vermis was involved, impairments included defects of planning, abstract reasoning, working memory, verbal fluency, visual–spatial organization, agrammatism and abnormal prosody, while lesions of the anterior lobe caused only minor changes in executive and visual–spatial functions. It may be that these findings are due to disruption of the cerebellar modulation of neural circuits that link prefrontal, posterior parietal, superior temporal, and limbic cortices with the cerebellum, due, in part, to diaschisis.

Complex orofacial movements can occur after resection of a cerebellar tumour. Van Mourik et al. [13] studied five children with such movements during the mute phase and shortly after the resumption of speech. Emotional phonation and chewing remained intact, and swallowing problems soon resolved, but the same movements could not be performed on request. As a result of their investigations, depression did not seem to account for these findings, nor did dysphasia, dyspraxia, or severe ataxic dysarthria, although the latter persisted after the resumption of speech. It is postulated that the mechanism involved may be a disruption of the initiation of intentional movements caused by cerebellar damage and by disturbed function in other cerebral areas, especially the frontal regions.

Apart from speech disorders, other neurological complications occur after surgery to constitute the posterior fossa syndrome, but unlike mutism, these are often not reversible. They include ataxia, tremor, cranial nerve palsies, bulbar palsies, hemiparesis, cognitive impairment, and emotional lability. The syndrome is most often associated with medulloblastomas, and sometimes with ependymomas or astrocytomas. A significant risk factor for the syndrome is involvement of the brain-stem, so that limitations to surgical removal of these tumours must be considered if this is possible [14].

4. Types and sites of the lesions

Erşahan et al. [15] reviewed 46 cases, including seven of their own. Their ages ranged from 2 to 61 years, but 91% of them were children. The cerebellar vermis was the site in more than 90%, and except for one arteriovenous malformation, these were large tumours of various kinds. The latency for the development of mutism ranged from 0 to 6 days, and lasted from 4 days to 4 months. After the transient mutism resolved, dysarthric speech ensued in 35 patients.

In the report of Koh et al. [16], the histories of six children were reviewed. Four had operations for cerebellar tumours, one had an arteriovenous malformation, and one had suffered from a head injury. In the majority of reports, the mutism appeared to be due to involvement of the dentate nucleus and its outflow dentatorubrothalamic tracts to the brain-stem, and to the cortex, in the superior cerebellar peduncle. The condition may have been exacerbated in some instances by hydrocephalus. Even retraction of the cerebellum may be sufficient to cause mutism [17], and transient mutism has been recorded among adults undergoing callosotomy for drug-resistant epilepsy which may have been due to surgical manipulation [18]. This suggests the delayed onset in some cases may be the result of vasospasm.

Other diagnoses that have to be considered are: a post-
ictal aphasia; a manifestation of basilar migraine, possibly without headache; the sudden onset of deafness, for example after bacterial meningitis; vascular lesions; the Landau–Kleffner syndrome; the autistic spectrum; and degenerative brain diseases.

Visual impairment has been associated with mutism after posterior fossa surgery in children. This may be due to a loss of cortical vision, as in the children reported by Liu et al. [19], the pupil reactions were normal, or may be the result of ischemia or spasm causing temporary dysfunction of the oculomotor nucleus that resolves with time. If this is suspected, neurophysiological and neuroradiological investigations may demonstrate involvement of the visual pathways [20].

5. Treatment

It is now accepted that emotional and physical trauma do not play a big role in causing elective mutism, and it is much more likely to be due to biologically mediated temperament and anxiety components. This is supported by a frequent family history of these traits, which may put the child at risk of similar problems [21]. This has affected the approach to treatment, but that is not to say that mutism does not follow a sudden dramatic episode. For example, Scabo [22] sights a girl of 5 years who became mute after her mother was murdered. Due to her age, she was initially not told about this, and when her mother did not appear she stopped talking, but when an explanation was given and she was taken to her mother’s grave, she started to speak again.

Treatment must concentrate on the training of basic social skills and activities of daily life so that the affected children can accept the company of others, and can express, at least in writing, their academic skills. The program of treatment will have to be targeted as much to the parents as to the child. This should stress the expectation that the child will talk again, encourage the use of all forms of communication, for example gestures, and all attempts at speech, such as whispering. All kinds of punishment and insistence on speech must be discouraged, like forcing the child into whispering. All kinds of punishment and insistence on speech, such as lorazepam and oxazepam have been shown to be effective in treating a girl with selective mutism and an obsessive–compulsive disorder with resolution of her speech disorder, but not of her other symptoms.

Another suggested treatment is with phenelzine, an irreversible non-selective monoamine inhibitor, in doses ranging from 30 to 60 mg/day lasting for 24–60 weeks. It seems to be well tolerated, weight-gain being the most common side-effect. There were no hypertensive reactions, or symptoms that might be related to serotonin effects, in the four children reported. It is especially beneficial when a weight-gain is desirable or the child suffers from enuresis. Also, co-morbid obsessive–compulsive symptoms, anxiety, irritability, and shyness may improve. However, because of food and drug interactions, it is probably best to reserve it for patients who have failed to respond to other treatments [31]. One girl has been reported, who suffered from elective mutism and Tourette’s syndrome, who did not respond to treatment with fluoxetine, fluvoxamine, or phenelzine, but apparently did to haloperidol [32], and benzodiazepines such as lorazepam and oxazepam have been shown to benefit psychomotor retardation and mutism [33], and diazepam in the case of an adult with epilepsy as well as lack of speech [34]. Also, bromocriptine, a dopamine agonist, has been suggested as monoaminergic pathways may be involved, possibly as a result of complicating hydrocephalus, and this has been effective in the treatment of one girl after surgery for a fourth ventricle choroid plexus papilloma [35].

If vascular spasm is a factor in causing the posterior fossa syndrome after surgery, and other lesions, in the cerebellum, it has been suggested that pretreatment with the calcium channel antagonist, nimodipine, might prevent the development of the disturbed blood flow due to vasospasm [36]. Also, although mutism following posterior fossa surgery may resolve after a short period of time, similar treatment as for the elective type, especially with drugs may be worth trying.

Psychotherapeutic approaches may well be unsuccessful [26], but there have been reports of beneficial responses to drug treatments. However, even if drugs have to be given, especially when mutism is combined with anxiety, difficult behaviour, and maybe learning difficulties, it is probably more important to enlist the aid of a team of experts, including speech therapists, occupational therapists, teachers, and psychologists [27].

There have been a number of favourable reports on the use of fluoxetine (prozac®), a selective serotonin reuptake inhibitor, in the treatment of selective mutism [28]. Dummit et al. [29] treated 21 such children, aged 5–14 years, with this drug in a dose ranging from 10 to 60 mg, and speech improved and anxiety lessened in 76% of cases. The trial lasted 9 weeks, but sometimes treatment had to be given for a longer period, and there were no controls. None of the children suffered from depression, so the improvement could not be attributed to the anti-depressant effects of the drug. Lafferty and Constantino [30] used fluvoxamine (Luvox) in treating a girl with selective mutism and an obsessive–compulsive disorder with resolution of her speech disorder, but not of her other symptoms.

As far as the child is concerned, behaviour therapy that offers rewards for increased, speech or attempts at speech, may be helpful, and one technique which has sometimes proved helpful is audio feedforward treatment. In this, audiotapes are edited of the affected child speaking in situations in which the child is currently not speaking. These are then listened to in these places [23]. Rye and Ullman [24] reported the successful long-term treatment of a boy with selective mutism with systematic desensitization, consultation with school personnel, and training in social speaking skills, and they stressed the importance of combining all three approaches. It may be especially important to establish a behavioural programme to reinforce any type of communication, non-verbal and then verbal [25].
6. Conclusions

Although elective mutism may be one end of a spectrum from shy children to those in whom there is a presumptive cause, such as immigration, unhappiness at school, or anxiety over some social disaster, it will be important to reserve management and treatment to those in whom the problem is significantly affecting the child, and or, the parents. Otherwise, as stressed by Kolvin and Fududis [37], a lot of children will be labelled unnecessarily who are reacting not unreasonably to a strange situation, such as a reception class in school, and who will soon be talking normally. These authors found the main characteristics of children with elective mutism were: immaturity of development, especially of speech, with an excess of speech abnormalities; an excess of behaviour problems, and a high incidence of enuresis and soiling; in most cases, a gradual development of shyness from early infancy; performance IQ covering most ranges of ability (verbal ability not being used as a measure of intelligence for obvious reasons), but there was a significant excess in the lower ranges; a high rate of psychiatric disturbances in the families of elective mute children; and the finding that the condition may be intractable.

If the condition is explained on the basis of extreme shyness and anxiety, parents, teachers, and others are more likely to understand and relate better to the children than if it is suggested that it is a question of defiance. This encourages a supportive, sympathetic, behavioural approach which can improve the child’s self-confidence and social interaction [31].

Mutism from organic causes is most frequently the result of posterior fossa surgery, usually to remove large tumours, and also after operations on the corpus callosum for intractable epilepsy [38]. The evidence shows that splitting the vermis does not of itself cause mutism, but that oedema or ischaemia resulting from vasospasm and involving the dentate nucleus and its connections may account for the condition, and its delayed onset and transient nature [39]. Also, hydrocephalus can be a contributing factor.

From the study of 26 children who had undergone surgery for the removal of cerebellar hemisphere or vermis tumours, it was concluded by Riva and Giorgi [40] that those with right cerebellar tumours presented with disturbances of auditory sequential memory and language processing, whereas those with left cerebellar tumours showed deficits on tests of spatial and visual sequential memory, and sometimes an impairment of prosodic intonation. Lesions in the vermis caused either mutism which could evolve into speech or language disorders similar to agrammatism, or behavioural disturbances ranging from irritability to behaviour similar to autism. This may mean that the cerebellum has a role in the modification of thought, language, and executive abilities, as well as social functions, and that this may operate early in childhood.

On recovery from mutism due to organic causes, the patient can often be dysarthric. However, as mentioned, this may not have the typical pattern associated with cerebellar involvement with staccato, slurred, jerky, forced, explosive, irregular, and most particularly, scanning speech, but speech may be monotonous and poorly articulated [10].

Studies with single photon emission tomography (SPECT) have shown that after surgery on the left side of the cerebellum for an astrocytoma, there was a marked reduction of cerebral perfusion in the right fronto-parietal region in one child, and in the left fronto-temporo-parietal area in another after removal of a medulloblastoma. When the patients regained normal speech, the SPECT returned to normal [41]. Similar findings have been shown in one patient by Erşahin [42], with the suggestion that this may be due to oedema or ischaemia. It may well be that techniques such as magnetic resonance imaging (MRI) and SPECT will explain the cause of mutism due to posterior fossa lesions.

References


