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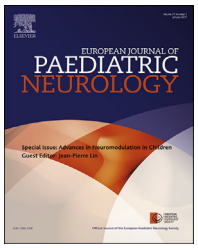

Original article

Bilateral globus pallidus internus deep brain stimulation for dyskinetic cerebral palsy supports success of cochlear implantation in a 5-year old ex-24 week preterm twin with absent cerebellar hemispheres

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ABSTRACT

Background: Early onset dystonia (dyskinesia) and deafness in childhood pose significant challenges for children and carers and are the cause of multiple disability. It is particularly tragic when the child cannot make use of early cochlear implantation (CI) technology to relieve deafness and improve language and communication, because severe cervical and truncal dystonia brushes off the magnetic amplifier behind the ears. Bilateral globus pallidus internus (GPi) deep brain stimulation (DBS) neuromodulation can reduce dyskinesia, thus supporting CI neuromodulation success.

Methods: We describe the importance of the order of dual neuromodulation surgery for dystonia and deafness. First with bilateral GPi DBS using a rechargeable ACTIVA-RC neurostimulator followed 5 months later by unilateral CI with a Harmony (BTE) Advanced Bionics Hi Res 90 K cochlear device. This double neuromodulation was performed in series in a 12.5 kg 5 year-old ex-24 week gestation-born twin without a cerebellum.

Results: Relief of dyskinesia enabled continuous use of the CI amplifier. Language understanding and communication improved. Dystonic storms abated. Tolerance of sitting increased with emergence of manual function. Status dystonicus ensued 10 days after ACTIVA-RC removal for infection-erosion at 3 years and 10 months. He required intensive care and DBS re-implantation 3 weeks later together with 8 months of hospital care. Today he is virtually back to the level of functioning before the DBS removal in 2012 and background medication continues to be slowly weaned.

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Conclusion: This case illustrates that early neuromodulation with DBS for dystonic cerebral palsy followed by CI for deafness is beneficial. Both should be considered early i.e. under the age of five years. The DBS should precede the CI to maximise dystonia reduction and thus benefits from CI. This requires close working between the paediatric DBS and CI services.

Deep brain stimulation (DBS) is a recognised neurosurgical intervention for a number of monosymptomatic genetic dystonias[1] and there are a growing number of reports of DBS benefits for dystonic cerebral palsy (DCP) which is the most common cause of childhood dystonia.[2–7] A detailed discussion of the benefits of DBS in DCP compared to genetic dystonias can be found elsewhere.[1,8–10] A major point of interest is the extent to which the dystonia of DCP can improve[11] and functional priorities can be met following DBS.[12–14] This is a particularly important issue since dystonia has a severe impact on the lives of children, without improvement in over 90% of cases and actually worsening in more than two thirds of cases, irrespective of aetiology.[15] Unfortunately most intervention studies for childhood dystonia do not address the concerns of children and their carers.[16]

This report describes the very special circumstance of DCP and sensorineural deafness requiring deep brain stimulation (DBS) and sensorineural deafness requiring cochlear implantation (CI): but when should these interventions be delivered and in what order?

1. Methods

In 2006 the Complex Motor Disorders Service (CMDS) was in an embryonic state of development with experience of 12 cases of DBS in children, mostly relating to neurodegeneration with brain iron accumulation (NBIA).[17] Our two cases of DYT-1 positive childhood dystonia had received DBS implants in Montpellier under Professor Philippe Coubes and his group in 2003 and 2004 respectively and one case of presumed genetic monogenetic dystonia (anarthric, NG fed and non-ambulant) was considering entering our DBS programme.

We were referred two 3-year old ex-24 week gestation-born premature twins, conceived via IVF, with severe 'motor spasms'. The twins, a boy and a girl, had severe dystonia also accompanied by hypotonia, most marked in the boy. The parents had not been made aware of 'dystonia' in relation to their children, so this was an additional diagnosis arising from the first neurology consultation. In addition the boy had suffered bilateral haemorrhagic infarction of the cerebellar hemispheres which were completely absent with only a little cerebellar vermis remaining. Both children had Gross Motor Function Classification System (GMFCS) and Manual Abilities Classification System (MACS) Level V.[18] Neither had made any significant motor progress (Videos 1 and 2). The boy (Fig. 1) was most severely affected, particularly with very severe nocturnal dystonia impairing sleep and periods of severe dystonia which the parents came to refer to as 'dystonic storms'. Today we would have classified the dystonia severity as DSAP level 3[19] i.e. unable to tolerate sitting comfortably, poor sleep patterns and a great risk of sliding into frank status dystonicus.[20] Both twins were orally fed but underweight and effectively experiencing 'faltering growth' or 'failure to thrive'. Fluid intake was a major issue for the boy, contributing to the formation of a large left renal calculus diagnosed in December 2007 requiring lithotripsy in April 2008. Two years of medical management of severe episodic dystonic crises with trihexyphenidyl, chloral hydrate at night for sleep and emergency diazepam during acute admissions for the boy ensued. At the age of five, the boy was confirmed to be completely deaf, although he had originally passed a 'new-born' hearing check. Although the exact mechanism of the progressive deafness remained unknown, cochlear implantation was discussed. However, concerns were raised that the severe rapid dystonia-choreoathetosis (dyskinesia) particularly causing dynamic neck and trunk forward flexion and slow rotational dystonic neck tremor would interfere with the cochlear implant magnet that usually sits over the cochlear implant behind the ear. The boy was hypotonic at complete rest, which is characteristic of DCP. Indeed his usual hearing aids were seldom in place, whistling continuously as they were dislodged from the ears by the dyskinetic head and neck movements against the wheel-chair head-rest (Video 3). A very important consideration for our DBS service was the decision to move exclusively to rechargeable DBS neurostimulator implantation with a 9 year battery life since our experience had shown that non-rechargeable batteries needed replacement every 18–24 months: a heavy burden for children and their families, with the added risks of device infection at each operation.[21] Given the need to proceed to cochlear implantation as soon as possible, the respective device companies were contacted to determine if there was any possibility of interference during the charging and the operation between the two neurostimulation devices: the Harmony (BTE) Advanced Bionics Hi Res 90 K cochlear device and the Medtronic ACTIVA RC rechargeable 16 channel neurostimulator newly released in Europe in the autumn of 2008. No obvious technical conflict between the devices could be established. Because the ACTIVA RC had never been used in an adult let alone a young child in the UK before, the surgery was agreed by the New Surgical Procedures Committee at King's College Hospital. In addition, the Complex Motor Disorders Service at the Evelina London Children's Hospital and the Cochlear Implant Services, both on the St Thomas' Hospital campus, met to discuss a shared strategy which included the order of surgery. It was agreed that DBS, requiring intraoperative in-frame MRI for stereotactic surgery should be performed first, followed 5