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Rechargeable Deep Brain Stimulators in the Management of Paediatric Dystonia: Well Tolerated with a Low Complication Rate

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Key Words
Dystonia · Pallidal stimulation · Deep brain stimulation · Battery

Abstract
Background: Deep brain stimulation (DBS) is a recognised method of treatment for primary and secondary dystonia. The size of non-rechargeable batteries has limited their use in small children. Our severe dystonia patients have required battery replacement every 20–24 months. ***Objectives:*** To evaluate reliability, care burden, patients' satisfaction and complications related to the rechargeable neurostimulator Activa® RC (launched by Medtronic in Europe in autumn 2008). ***Methods:*** Complications were recorded prospectively, and a questionnaire on neurostimulator maintenance, care burden and parental satisfaction was applied to all patients with at least 3 months of follow-up. ***Results:*** 30 Activa RCs were implanted between December 2008 and June 2010, 25 with a follow-up of 3–17 months (mean 10); the mean patient's age at surgery was 11.1 years; 22/25 questionnaires were completed. All families achieved good standards of recharging. Caregivers were responsible for recharging in 82% of cases. With higher parameters of stimulation, recharging time was longer than initially recommended by the manufacturer. All but one family would recommend Activa RC to other patients. Transient recharging problems were the most common complication (36% of cases). Infection/skin erosion occurred in 8% of cases, self-resolving early seroma in 20%. ***Conclusions:*** Activa was found to offer reliable stimulation with a low rate of significant complications and a suitable treatment option for children with dystonia.

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Introduction

Deep brain stimulation (DBS) is a well-recognised method of treatment for dystonia in adults and children [1, 2]. Initially used for patients with Parkinson's disease and idiopathic dystonia, DBS has recently been helpful in treating patients with secondary dystonia [3, 4]. Childhood dystonia may be a severe and debilitating condition, frequently of secondary aetiology, where treatment with medication alone often proves unsuccessful.

Non-rechargeable implantable neurostimulators such as the single-channel Soletra® and double-channel Kinetra® neurostimulators (Medtronic Inc., Minneapolis,

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	Activa RC	Activa PC	Kinetra	Soletra
Volume	22 cm^3	39 cm^3	50 cm^3	26 cm^3
Weight	40 g	66 g	82 g	42 g
Case thickness	9 mm	15 mm	15.2 mm	10 mm



Fig. 1. Comparison of sizes between different types of neurostimulators used in our centre. Information provided by Medtronic Inc.

Minn., USA) which have been the mainstay of DBS practice have several disadvantages. Dystonic children are frequently small, underweight due to high-energy requirements and feeding problems. Neurostimulator device size (fig. 1) may limit the number of children suitable for DBS. Two Soletra single-channel stimulators, required for bilateral pallidal stimulation, are implanted a minimum distance of 20 cm apart. This increases both risks of infection and surgical time. Although a single Kinetra stimulator provides bilateral stimulation, the size limits its use in younger children under 30 kg in weight. Both Soletra and Kinetra stimulators may unexpectedly switch off leading to therapy disruption [5, 6], and the battery end-of-life phase is highly clinically disruptive to patient well-being.

For patients with Parkinson's disease the reported mean longevity of implantable non-rechargeable stimulators is 37–45 months [6, 7]. The use of monopolar stimulation, high voltage, wide pulse width of 150–450 μs and frequency of stimulation in the 130- to 210-Hz range leads to higher energy consumption and shorter battery life in cases of secondary or primary dystonia [3, 8–11]. We have found a battery longevity between 15 and 33 months (mean 24 months) and observed reduced control of symptoms even prior to neurostimulator end of life, particularly in secondary dystonias which were often the severest cases [11]. This results in multiple surgical battery replacement procedures. A child receiving his/her first DBS implant at the age of 7 years could conceivably require 4–9 non-rechargeable battery replacements before reaching adulthood.

Given the relatively short non-rechargeable battery life, the rechargeable neurostimulator Activa® RC (Medtronic Inc.) potentially offers the prospect of fewer disruptions to dystonia management from end-of-life battery failure, fewer operations to replace batteries, a reduced operative and postoperative complication rate, fewer general anaesthetics, hospital admissions and psychological stresses.

Due to its small 22-cm^3 volume, it has been possible in our service to offer DBS to children weighing as little as 11.5 kg. One drawback of rechargeable batteries is the need for regular recharging, requiring thorough individual teaching and practical training sessions in the preoperative and the postoperative periods for patients and caregivers. There is also the potential for problems arising from poor caregiver compliance. We aimed to evaluate device reliability, care burden, caregiver satisfaction and technical and clinical complications related to the Activa RC deep brain stimulator.

Methods

Surgery

All DBS surgeries were performed under general anaesthesia as a one-stage procedure. A Leksell frame was used in all cases, finger tightening the screws in the very young patients. DBS leads (model 3389, Medtronic) were implanted bilaterally, through frontal incisions and burr holes, into the posteromedial globus pallidus. All new DBS patients underwent direct anatomical targeting using pre-operative high-resolution proton density and T_1-weighted volume stereotactic brain MRI scans. Medtronic Framelink software was used to calculate the coordinates of the targets. A postoperative stereotactic CT scan was performed in all patients to confirm optimal electrode placement. Intra-operative impedance or micro-electrode recordings were also used to help localisation of the target. Patients with de novo implants received a 7-day prophylactic course of intravenous antibiotic.

For battery replacement procedures with an Activa RC, 24–48 h of intravenous antibiotic prophylaxis was used peri-operatively. An adaptor was used for connection of the existing cables, implanted with the original Soletra and Kinetra batteries, to the new Activa RC.

Over a period of 17 months, between December 2008 and June 2010, all cases with implanted Activa RC neurostimulators were monitored prospectively for possible complications. Cases with follow-ups longer than 3 months were entered for further analysis.

All families (children and caregivers where relevant) received information sessions on recharging the stimulator prior to surgery and individual practical training during the 7 postoperative days of inpatient admission until they felt confident with the Activa RC DBS stimulator maintenance. Families were advised to recharge the DBS stimulator initially daily to ensure the process became a routine care activity to maintain a high battery charge of 75–100%.

A questionnaire on maintenance, additional care burden and user satisfaction was given to all caregivers of children with follow-ups longer than 3 months (25 cases). Ten questions were

asked, relating to recharging frequency, method, time required, care burden and user satisfaction. Questionnaires were completed during routine follow-up appointments, over the phone or returned via e-mail.

Results

Thirty Activa RC neurostimulators were implanted between December 2008 and June 2010: 20 de novo implants and 10 battery replacements arising from previous battery end of life.

Twenty-five children and young people with a follow-up of at least 3 months were identified. The mean age at Activa RC insertion was 11.1 years (range 4.2–19 years). Causes of dystonia included 5 patients with primary dystonias (2 cases DYT1 positive and 3 non-DYT1) and 20 cases with secondary dystonias of different underlying causes. Demographic data is presented in table 1.

The mean follow-up was 10 months (range 3–17 months).

User's Questionnaire

The user's questionnaire (fig. 2) was given to caregivers of 25 patients (15 new DBS patients and 10 replacements) with a minimum follow-up of 3 months.

Twenty-two out of 25 (88%) questionnaires were returned: 16 questionnaires were completed during routine follow-up appointments, 4 over the phone and 2 were returned via e-mail.

Responsibility for Recharging

Caregivers recharged the stimulator in 18/22 (82%) cases. However, 4 children or young people were responsible for recharging (those with good manual function, independent mobility and good intellectual function). The youngest 'self-charger' was a 7-year-old girl with DYT1-positive dystonia.

Frequency of Recharging

Nineteen out of 22 stimulators (86%) were recharged at least once every 3 days, and 10 were recharged daily.

Duration of Recharging

In 12/22 (54%) cases, the Activa RC stimulator was charged for up to 30 min during a single session and in 18/22 (82%) cases for up to 60 min. Charging time was inversely proportional to the frequency of sessions, i.e. a shorter time was required if sessions took place daily or on alternate days.

Batteries were charged to at least 75% capacity at the end of each session in 20/22 (91%) cases. A significant proportion of caregivers chose to recharge the stimulator during the child's sleep (9/22, 41%), while 13/22 caregivers (59%) used quiet activity time such as watching TV, playing computer games or tea time to recharge the Activa RC.

Usefulness of the Recharging Harness

Only 5/22 patients (23%) used the harness provided with the stimulator to maintain coupling between the antenna and stimulator during every session. Thirteen out of 22 (59%) families found the harness unhelpful due to the small size of their child, residual dystonic/dyskinetic movements or when recharging during the child's sleep. Alternatively, the recharger's antenna was secured to clothing with tape or held by the caregiver for the required time.

Maintaining Contact (Coupling) during Recharging

No problems were found in maintaining a good connection between the antenna of a recharger and the Activa RC stimulator (coupling) in the majority of cases (15/22, 68%). However, some difficulty was reported in 7/22 cases (32%).

Care Burden of Recharging

Nineteen out of 22 (86%) caregivers/patients did not consider recharging as a significant care burden, whilst only 1/22 (4%) would not recommend a rechargeable stimulator to other patients. Reasons stated in favour of the Activa RC by families included its smaller size and the prospect of not requiring a battery replacement for 9 years.

Hardware-Related Complications

Nine out of 25 (36%) patients experienced temporary difficulty with recharging the stimulator. In 3 (12%) of these cases, the recharging set required replacement without risk to the children (table 1).

In 2 (8%) cases for whom a previous Kinetra stimulator was replaced with an Activa RC, the adaptor moved spontaneously in front of the Activa RC stimulator leading to ineffective recharging. The problem resolved following external manipulation of the system without the need for surgical revision. We identified poor compliance with recharging recommendations in 4/25 (16%) patients during the first 3–6 months resulting in low battery voltage in all cases and disruption of stimulation in 2 cases. Additional teaching and more frequent reviews to ensure better compliance resolved the problem in all 4 cases.

Table 1. Demographic data and hardware-related complications

Patient No.	Sex	Diagnosis	Age at Activa RC implantation, years	Length of follow-up months	New DBS implant/battery replacement	Complications
1	M	DYT1	14.1	5	New	
2	F	DYT1	7.3	16	New	
3	F	Idiopathic dystonia; non-DYT1	17.4	3	New	
4	M	Idiopathic dystonia; non-DYT1	13	10	Replacement	
5	F	Idiopathic dystonia + learning difficulties; non-DYT1	15.3	6	Replacement	Problems with recharging at 3 months, adaptor in front of the stimulator; resolved after external manipulation
6	F	Ex-preemie CP	5.5	15	New	
7	M	Ex-preemie CP	6.0	8	New	Early seroma – resolved Problems with recharger; recharger replaced
8	M	Ex-preemie CP	5.3	17	New	Early seroma – resolved
9	M	Kernicterus-CP	12.3	14	New	Battery migrated 11 months after implantation; battery repositioned, skin erosion necessitating battery removal; awaiting re-implantation
10	M	Kernicterus-CP	12.0	15	New	Problems with recharger; recharger replaced
11	M	Ex-preemie CP	5.4	13	New	
12	M	Kernicterus-CP	9.7	7	New	Battery flat at 2/12 due to poor compliance with recharging; resolved after additional training
13	F	Dystonic CP	12.1	6	New	Recharging problems within first 6 months – poor compliance; resolved with additional training
14	F	Dystonic CP	6.4	11	Replacement	Stimulator switched off by carers incidentally Recharging problems – poor compliance – first 6 months; resolved with additional training
15	F	Dystonic CP	6.0	8	New	
16	M	GA type 1	11.7	6	New	Early seroma – resolved Problems with recharger; recharger replaced
17[1]	M	Hemidystonia-CVA	13.9	6	New	Initial compliance problem – resolved with additional training
1[2]	F	Hemidystonia-CVA	18.9	12	Second replacement	Problems with recharging at 8 months – adaptor displaced at the front of stimulator; resolved after external manipulation
19	F	Dystonia, polymicrogyria, epilepsy, atypical MECP2 mutation	15.5	4	Replacement	
20	F	NBIA-PANK2	19	10	Replacement	Infected within 1/12; stimulator and extension wires removed; awaiting re-implantation
21	M	NBIA-PANK2	4.2	8	New	
22	F	NBIA-PANK2	14.2	6	Second replacement	
23	F	NBIA	10.1	12	New	
24	F	NBIA-PANK2	10.1	15	Second replacement	Early seroma – resolved Stimulator switched off accidentally by family at 11 months – severe status dystonicus – resolved after switching back on
25	M	NBIA-PANK2	12.5	14	Replacement	

DYT1 = Primary torsion dystonia with positive DYT1 gene mutation; CP = cerebral palsy; GA = glutaric acidaemia; CVA = cerebrovascular accident; NBIA-PANK2 = neuronal degeneration with brain iron accumulation associated with mutations in the pantothenate-kinase gene.

[1] Patient receiving unilateral DBS to the globus pallidus internus.

[2] Patient receiving unilateral DBS to the globus pallidus internus and nucleus ventralis oralis anterior of the thalamus.

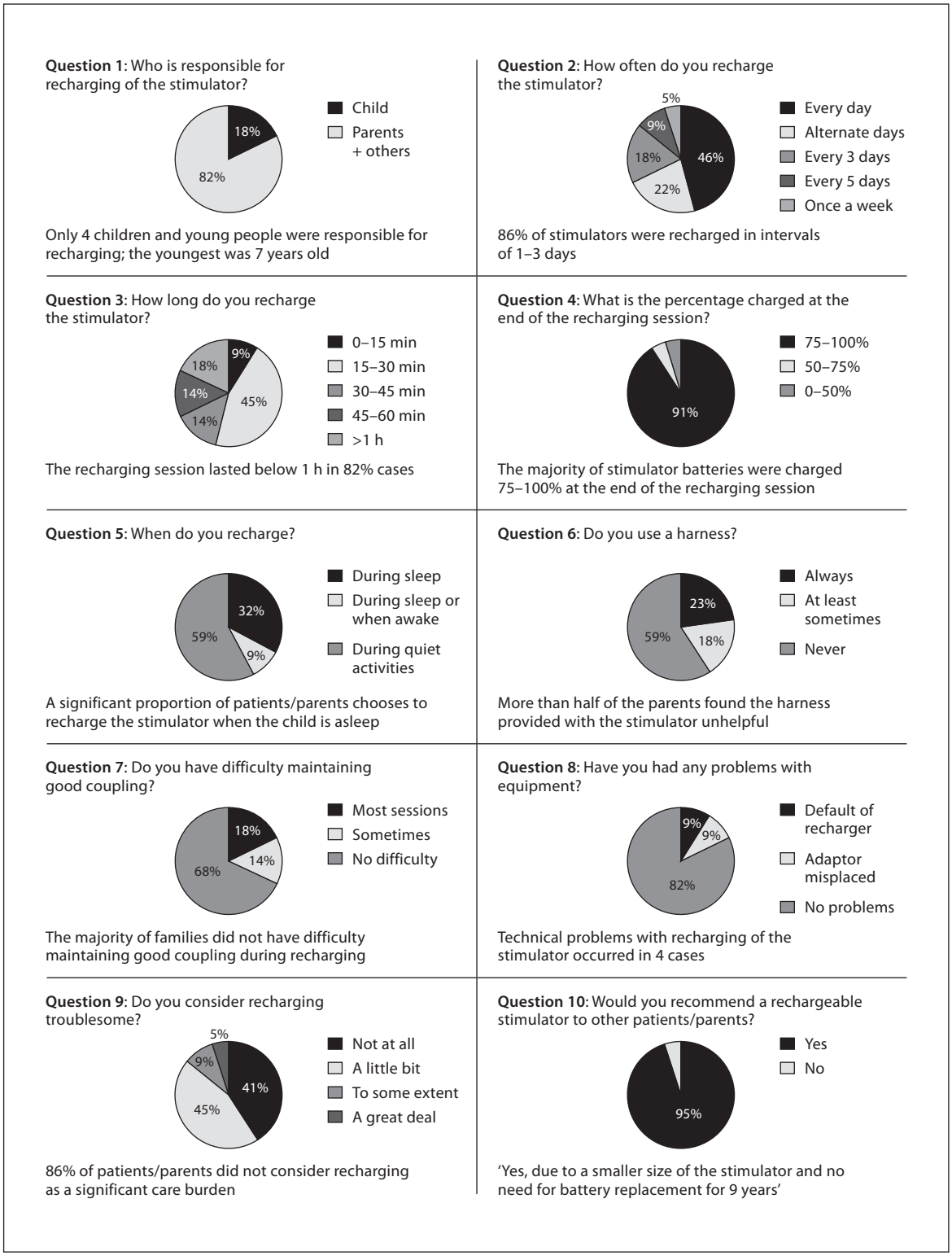

Fig. 2. Results of the questionnaire on Activa RC maintenance and parental satisfaction.

Two out of 25 (8%) stimulators were accidentally switched off by caregivers. Additional training was given, following which there was no recurrence of this problem.

One of 25 (4%) stimulator batteries became infected within the first 2 weeks following implantation and was refractory to treatment with intravenous antibiotics resulting in partial system removal. The intracranial parts remain in situ, and the patient is scheduled for re-implantation after a 6-month infection-free period of observation.

One of 25 (4%) stimulators migrated over the sternum towards the end of the first year of implant use and required surgical repositioning more laterally. This patient developed skin erosion over the Activa RC battery site which failed to heal and required eventual removal of the battery to allow skin healing.

Five of 25 (20%) cases developed seroma within 2 weeks of implantation, all resolving spontaneously.

Discussion and Conclusions

This is the first report to date on the experience with a rechargeable DBS neurostimulator in children and adolescents. Limitations of this study relate to the small numbers of patients, short follow-up and the use of a non-validated questionnaire. A longer observation period is required.

Overall the families appear to have incorporated Activa RC DBS stimulator recharging into their daily routines and did not consider it as a significant care burden. The majority of caregivers adhered to our recommendations for regular recharging sessions and maintained a high battery charge percentage. However, 4/22 (16%) of the cases required additional training and more frequent follow-up appointments to ensure good compliance. This indicates the importance of an appropriate patient/caregiver selection and training when offering a rechargeable neurostimulator for children. The need for additional professional input to support the families has to be incorporated into the DBS service work plan.

We were disappointed that the harness provided with the stimulator could not be used effectively in many of our small patients and together with the families had to work creatively to find alternative ways of securing the recharging antenna in the optimal place.

All but 1 family would recommend a rechargeable stimulator to other patients due to its smaller size and battery longevity of 9 years and therefore the reduced need for frequent battery replacement. Overall, the responders commented that the stimulator was not prone to being switched off. Those with experience of non-rechargeable DBS devices felt more in control by having the possibility to check the Activa RC stimulator's status with the patient control device. However, this led to the stimulator accidentally being switched off by the caregivers in 2 cases. When replacing the significantly larger Kinetra with the Activa RC, caution should be taken to avoid displacement of the adaptor anterior to the battery, possibly by suturing, to avoid problems with recharging.

Between May 2005 and December 2008, we had implanted 31 single-channel Soletra and 27 double-channel Kinetra neurostimulators in 32 patients. By June 2010, 45% of Soletra and 37% of Kinetra batteries were depleted. The infection rates were 10 and 11%, respectively. Unexpected switching off occurred at least once in 22% of Soletra and 11% of Kinetra stimulators. There was 1 case of seroma, 1 clinically and radiologically insignificant intracranial bleed at the time of surgery, 1 faulty electrode and 3 short leads requiring intervention.

The small size of the rechargeable neurostimulator offers the possibility of bilateral pallidal DBS in younger children. It has been reported that shorter disease duration prior to DBS correlates positively with achieved outcomes [3, 10].

In conclusion, the Activa RC offers a reliable and well-tolerated method of stimulation with a low rate of complications and thus is a suitable option for children with dystonia. However, with higher parameters of stimulation, recharging time is longer than initially recommended by the manufacturer. A smaller size harness or other means to maintain connection coupling during recharging would be useful for children. Long-term observation is required to monitor reliability of the rechargeable stimulator, care burden and adverse effects.

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Disclosure Statement

No conflict of interest.

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