DBS IN A THREE YEAR OLD CHILD WITH SECONDARY DYSTONIA DUE TO NEONATAL HYPERBILIRUBINEMIA

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DISCLOSURES

NO DISCLOSURES
INTRODUCTION

Hyperbilirubinemia is one of the most common causes of readmission in the early neonatal period.

In some cases prolonged high levels of bilirubin lead to severe neurotoxicity (0.4 – 2.7/100 000 births in the western hemisphere).

Damages in the basal ganglia lead to severe neurological deterioration (Dystonia, spasticity, mental retardation); Fig. 1.

Deep brain stimulation (DBS) of the internal pallidum (GPI) can improve dystonic symptoms. Especially in primary generalized dystonia, results in secondary dystonic movement disorders are varying.
METHODS

We report on a three year old girl with severe bilirubinencephalopathia.

She was born mature and in due time without complications.

On day one she developed jaundice, 2 weeks later she showed tonic movements.

Increase of symptoms with lack of motor development, she showed random untargeted dystonic and opisthotonic movements, not able to control head and trunk.

Medication with dopamin and vigabatrin showed no effect. Clobazam alleviated symptoms but caused distinct sedation.
METHODS

After intensive ethical discussion with all involved medical fields, the bilateral implantation of DBS-leads in the GPI was indicated. The parents signed informed consent.

Under general anesthesia a stereotactic frame (Inomed ZD-System) was fixed to the head and a stereotactic MRI-scan performed (T₁ MPR+Gad. and T₁ IR) Fig. 2.

Under general anesthesia the leads (Medtronic 3389-28cm) were implanted using microelectrode recordings and intraoperative test stimulation. The electrodes were connected to a Medtronic Activa RC IPG which was implanted paraumbilically. Surgery was an „all-in-one-procedure“.
RESULTS

The operation had no complications, a postoperative CT-Scan showed the leads localized in the prior planned target (GPI)

The patient showed a minor microlesion effect

First stimulation parameters: G+; 9-; 1V; 60µs; 130Hz and G+;1-; 1V; 60µs; 130Hz.

Over the course of time stimulation was increased to 2,5V bilaterally

The symptoms improved, especially the opisthotonotic phases decreased significantly, swallowing became possible
RESULTS

Figure 1:
RESULTS

Figure 2:

Trajectories for the GPI on the right (green) and the left side (red). Screenshots from the planning software brainlab iPlan Vs 3.0
DISCUSSION

In our case the patient improved under stimulation therapy.

One of the youngest DBS-patients ever reported.

In patients with primary generalized dystonia the stimulation effect is remarkable what is verified in many trials.

In many cases GPI-DBS fails to improve symptoms in secondary dystonia like cerebral palsy or dystonic movement disorders in patients with NBIA.
SUMMARY POINTS

The implantation of DBS electrodes is a safe procedure and can be performed even in very young children.

The indication has to be proved intensively and all risks and ethical aspects have to be discussed with all participating medical disciplines and the patient and his/her family prior to surgery.

When there is no other treatment opportunity than deep sedation, the implantation of DBS-electrodes to the GPI should be carefully considered.