Letter to the editor

Clinical practices in intrathecal baclofen pump implantation in children with cerebral palsy in France

A R T I C L E  I N F O

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Dear Editor,

Cerebral palsy (CP) encompasses an array of permanent movement and posture disorders leading to activity limitations caused by events or non-progressive pathologies affecting the developing brain. In CP, motor disorders are often associated with sensory, perception, cognitive, communication and behavioral disorders, epilepsy and secondary musculoskeletal problems [1]. One of the commonly used therapeutics in CP is baclofen [2], which inhibits presynaptic and excitatory neurotransmitters [2]. The 2009 guidelines of the French healthcare safety agency Agence Nationale de Sécurité du Medicament recommend intrathecal baclofen (ITB) therapy via pump infusion for patients with widely distributed spasticity of the lower limbs diffusing to the trunk (Grade A recommendation) [3]. Studies with a high level of scientific evidence demonstrated that ITB was the most effective treatment for spasticity in CP patients with Gross Motor Function Classification System (GMFCS) grades IV and V [2,4]. A European consensus [5] has been established on the appropriate use of ITB mainly for non-ambulatory children with CP (GMFCS grade IV or V) [2,4]. However, in France, because of the great heterogeneity of patient conditions and medical practices, no standard of care has been established for ITB therapy.

We performed a survey of the current use of ITB via pump infusion in children seen in pediatric physical medicine and rehabilitation (PMR) departments in France to describe practices and to establish specific guidelines. We also surveyed the literature for practices.

The evaluation involved a questionnaire survey about clinical practices, specifically pump implantation and follow-up after ITB, with 108 closed questions regarding full management of ITB from patient selection to post-implantation follow-up. In May 2012, the questionnaire was sent by email to 29 heads of pediatric PMR departments all over France to be distributed via the French society for research in patients with disabilities (SFERHE) network to all physicians (PMR physicians, child neurologists) and surgeons (orthopedic surgeons, neurosurgeons) involved in administering ITB by pump infusion.

The response rate was high (27/29 centres): 21 centres delivered ITB by pump infusion. We received 24 questionnaires completed by 24 professionals, mainly PMR physicians (n = 17), but also neurosurgeons (n = 5) and orthopedic surgeons (n = 2). A total of 20/24 professionals worked in a public hospital. Patients were referred mainly by PMR physicians [23] but also child neurologists and neurosurgeons, most often after a multidisciplinary decision (20/24). The activity rate ranged from 5 to 20 patients/year.

For 23/24 teams, children with CP who benefited from ITB pump implantation had GMFCS level V. For 19/24 teams, children received oral baclofen before the team decided to implant the ITB pump.

The process of ITB pump placement involves 5 phases:

- refining indications and objectives for ITB pump placement;
- deciding on a valid outcome measure;
- testing the potential effectiveness of ITB pump placement with a pre-implantation trial;
- actual pump implantation;
- postoperative monitoring.

The indications and objectives for ITB pump placement need to be precisely refined [5], as was reported by 21/24 teams. According to Hoving et al. [4], the child must be older than 4 years old without any weight restriction, but in our survey, we found high variability in terms of weight limits for implantation. Hoving et al. [4] highlighted the need for a multidisciplinary discussion in the final decision for pump placement, as was reported by most of our professionals (20/24). Decisions against pump placement were based on age (5/24), body weight (15/24), skin abnormalities (11/24), socio-environmental issues (12/24), presence of gastrostomy tube (2/24), severe scoliosis (1/24) or spinal fusion (1/24). Sometimes parents refused ITB pump placement before the pre-implantation trial (14/24). Using validated tools in this population to highlight the efficacy of the pump during the test phase could standardize the pre-implantation evaluation. Spasticity must be assessed before implantation [4] and overall was properly conducted by all of our teams (16/24), most using the Ashworth scale (15/24).

Once objectives are set, a valid outcome measure such as the Goal Attainment Scale is recommended and should be developed in clinical settings and for research [6]. Only 5 centres used the Goal Attainment Scale to define and refine objectives with the family and patient. Other types of evaluations performed before the pre-implantation trial included video recordings of functional ability (5/24) and evaluation of pain (17/24), abnormal movements (15/24), contractures (12/24) and urodynamics. In all cases, this comprehensive assessment was conducted with the family and/or hospital team and/or referring medical and healthcare professionals. Overall, 21/24 centres qualitatively evaluated activity and

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participation, mainly by physicians during consultation, but they did not use adequate quantitative scales.

Once pump implantation is decided, the potential effectiveness of ITB pump placement needs to be evaluated with a pre-implantation trial, which was performed by most of our professionals (20/24). This trial predicts to some extent the expected outcomes for children, their family and healthcare professionals in cases of pure spasticity or mixed dystonia and spastic affections. The pre-implantation trial allowed for refining the objectives of ITB implantation (12/24), discussing the benefit/risk ratio (12/24) and involving the family in the first decision (13/24). Regarding practical modalities and according to the literature, a pre-implantation trial can consist of evaluating a single dose (=bolus) via lumbar puncture [12], eventually repeated while progressively increasing the dose [2]. As well, continuous perfusion with dose titration via an external catheter can be used [7]. In our survey, 15/24 physicians chose lumbar puncture for the pre-implantation trial. Standardizing the trial technique seems difficult, because each professional received different training and adopted specific habits. Lumbar puncture consisted in 1 to 3 boluses for 11 teams and was described as easy (13/24), fast (12/24) and safe (10/24). Other techniques such as an implantable chamber (12/24) and external catheter (8/24) are more complex, and there is great variability in the technique itself and also duration of use and number of tests to be performed. In 46% of cases, baclofen was not discontinued orally during the trial period.

Mean implantation delay after the pre-implantation trial ranged from a couple of days (7/24) or weeks (11/24) to months (5/24) and varied for 10 responders. The time to postoperative activation of the ITB pump varied greatly: immediately after the priming bolus (4/24) and between 2 and 4 days (15/24) and up to 28 days (3/24) after placement. The pump implantation was subcutaneous (19/24) and/or subfascial (9/24). Regarding the implantation site, some authors preferred subfascial pump implantation to decrease the risk of cerebrospinal fluid leaks [8]. Guidelines from 2010 [5] also recommended the subfascial location on the right side. Furthermore, the presence of gastrostomy in the upper-left abdominal quadrant was not a contraindication, nor was spinal arthrodesis for the insertion of an intrathecal catheter [3,5]. Preoperative examinations were rarely systematic and varied by the teams’ habits. Data from the literature yield little information on these examinations for daily clinical practice. A radiographic image for reference after pump implantation is recommended to monitor the progression of potential static disorders after baclofen-induced changes in muscle tone.

Postoperative precautions and follow-up modalities were rarely detailed. A compressive bandage was rarely indicated in the literature. In our study, patient progress after pump implantation differed by the various possibilities available in each structure. Specific instructions were given to clinical teams after surgery but not systematically (11/24): for the day when the patient is authorized to get out of bed (10/24), systematic application of a compressive bandage (6/24) or other types of instructions (4/24). Post-implantation monitoring included radiographic check-up of the pump and catheter placement in 7/24 of cases, investigating the pump [6] and other types of monitoring (1/24). In the immediate postoperative period, 16/24 of cases had cutaneous monitoring. The follow-up was based on investigating the pump (15/24), cutaneous monitoring (14/24) and more rarely, radiographic monitoring of the pump system (2/24). According to Hoving et al. [4], the follow-up should start early and continue without any interruptions. In our study, 5/24 of professionals reported delayed complications: 17/24 of physicians performed delayed post-implantation evaluation, most often when filling up the pump (15/24). After a long-term follow-up, 2/3 teams reported a positive effect for all cases. Recommendations from 2010 [5] help physiotherapists by providing advice, instructions on monitoring and pump-filling frequency.

Improved knowledge of the family and healthcare providers in terms of potential pump-related complications would improve ITB pump outcomes by preventing severe complications. A change in the patient’s clinical state should alert family and clinicians to potential complications or pump dysfunction. In accordance with the literature, most post-implantation complications could be prevented [5]. During the postoperative period, 16 teams reported complications, with no adverse events with the pump itself (only 1/24 reported adverse events). The most common complications were local infection (12/24 cases). Authors proposed subfascial implantation to try to decrease this complication, along with a preoperative course of prophylactic antibiotic therapy [9]. Another complication was catheter disunion (13/24 occurring “sometimes”) and this appeared in the literature, along with cerebrospinal fluid leaks (4/24), catheter dysfunction (3/24) [9] and onset of subcutaneous effusion (12/24 occurring “sometimes”), which suggests the relevance of applying a compressive bandage with the slightest doubt or even systematically. In the late postoperative phase, mechanical complications were the most common. These material-related issues should be discussed with the manufacturer; complications were frequent (5/24 in our survey).

The sudden ending of treatment could lead to withdrawal syndrome, which could become serious. Weaning risks are increased with intrathecal delivery; withdrawal syndrome diagnosis is even more difficult in patients with multiple disabilities, who are unable to express their discomfort. It can occur during catheter issues or sudden and unpredicted pump malfunction [10,11]. It was rarely mentioned in our survey. Professionals should know their patients and their specific care management well and should warn the family of telltale symptoms, especially when patients are travelling. Precise information should be given to patients and families for recognizing symptoms and reacting quickly by consulting a medical team in an emergency situation.

Managing spasticity in children with CP by ITB pump placement is an effective but demanding treatment. There is important heterogeneity in terms of evaluating treatment objectives, pre-implantation tests, implantation techniques and long-term follow-up. Expert teams working on a consensus draft based on current literature data could lead to the creation of a “Notebook” to precisely refine treatment objectives and their evaluation at different stages, define modalities for a pre-implantation trial, and list symptoms and signs suggesting pump malfunction or treatment complications. This logbook could help standardize pump implantation practices in accordance with international recommendations.

Disclosure of interest

The authors declare that they have no competing interest.

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References

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