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Review article

The neurosurgical treatment of patients in dystonic state – Overview of the literature

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ABSTRACT

Treatment options for patients in dystonic state include sedation, artificial ventilation, intrathecal baclofen infusions and stereotactic procedures. The main aim of this overview is the presentation and assessment of stereotactic procedures applied for treating patients in severe dystonic state. We performed literature overview starting from 1998 to 2012 with case reports regarding all patients treated by stereotactic procedures for dystonic state. We were able to find 15 articles describing 22 patients. Ablative procedures were described in 5 articles (3 thalamotomies, 3 pallidotomies) and were done in 6 patients. In the remaining 10 articles, globus pallidus internus stimulation was utilized in another 16 patients. We can conclude that bilateral pallidal deep brain stimulation seems to be the best stereotactic target for patients in dystonic state.

1. Background

Status dystonicus (SD), also known as dystonic storm, is a sudden and persistent episode characterized by acute worsening of dystonic movements, usually affecting patients with an established diagnosis of primary or secondary generalized dystonia. In view of its associated complications, status dystonicus is regarded as a life-threatening condition. There are several types of secondary dystonia which may be complicated by SD, including post-traumatic and post-inflammatory dystonia, and rapid exacerbation of dystonic movements in patients suffering from cerebral palsy. Certain neurodegenerative diseases such as Wilson disease or neurodegeneration with brain iron accumulation (NBIA, formerly called Hallervorden–Spatz syndrome) may also precipitate status dystonicus [1]. Trigger factors for SD typically include febrile infections, dehydration, dyselectrolytaemia, administration of medications (e.g. penicillamine in Wilson disease) and sudden discontinuation of drugs (e.g. anticholinergics, tetrabenazine, intrathecal baclofen or lithium).

Clinical symptoms of status dystonicus were first described by Jankovic and Penn in 1982 [2]. The first reported case was that of an 8-year-old boy with primary generalized dystonia whose neurological condition deteriorated significantly over a period of six months. The patient required sedation, paralysis and mechanical ventilation. Dystonic storm was managed

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with levodopa-containing drugs (Sinemet). Two years later, Marsden et al. [3] described two other patients with primary generalized dystonia who presented with a sudden-onset worsening of the disease. The condition of the patients was defined as ''desperate dystonics''. One of the patients, a 12-year-old boy, required sedation, paralysis and mechanical ventilation for two weeks. Pharmacological treatment introduced by the author (benzhexol, pimozide and tetrabenazine) proved effective and resulted in an improvement of symptoms. The other patient, a 15-year-old boy with dysphagia and frequent episodes of painful generalized dystonic spasms, however, failed to respond to the same pharmacological regimen. Pimozide was even found to increase the symptoms of dystonia in that patient [3]. Another case of effectively managed SD was reported by Narayan et al. [4] in 1991. It was a case of an 18-year-old man with predominantly axial dystonia caused by perinatal injury. The patient's condition deteriorated after spinal surgery, when opisthotonus and dystonic spasms of the lower extremities, abdominal and respiratory muscles were observed. The patient required sedation and mechanical ventilation. Pharmacological therapy with anticholinergic agents, tetrabenazine and oral baclofen was ineffective. In order to alleviate excessive muscle rigidity, the authors used an intrathecal infusion of baclofen which helped to achieve a nearly complete control of dystonia. Another report, authored by Vaamonde et al. [5], focused on two cases of status dystonicus in children. The authors were the first to apply the term ''dystonic storm'' to refer to the observed symptoms. One of the patients suffered from primary generalized dystonia and the other from NBIA. Both had to be sedated and mechanically ventilated. Pharmacological treatment was introduced; however, it proved ineffective in the patient with primary dystonia, while the second patient, who had presented with secondary dystonia, responded only to oral baclofen.

The historical overview presented above encompasses descriptions of isolated cases of SD. Subsequent years saw the publication of a few reports on a series of cases. The first authors to describe, in 1998, a group of 12 patients with SD episodes were Manji et al. [1]. The authors reviewed cases of SD covering a nearly 10-year period, and were the first to report on thalamotomies performed in SD patients. In one patient, bilateral thalamotomy brought no therapeutic effect. In another patient, who suffered from hemidystonia, however, the procedure contributed to a regression of SD. The largest group of patients with SD episodes was reviewed by Fasano et al. [6] in a retrospective multi-centre study covering a total of 89 episodes of SD which occurred in 68 patients over a span of nearly 18 years. The authors conclude that surgical treatment (globus pallidus stimulation) has proven to be superior to all other therapeutic modalities available for SD episodes.

Although SD is a rare complication of dystonia, it is a life-threatening condition. An appropriate diagnosis and rapid management are thus vital for patient survival. Current effective treatment modalities for SD include stereotactic implantation of electrodes in the internal globus pallidus. Paradoxically, however, sudden interruption of neurostimulation may also result in the development of SD.

2. Conservative (pharmacological) management of status dystonicus

Pharmacological treatment of primary and secondary dystonias remains an empiric therapy. The prevailing opinion is that the treatment of choice in children suffering from dystonia is based on levodopa. In dopa-responsive dystonia (DRD), the treatment results in complete regression of symptoms [1]. In order to confirm or rule out DRD, a therapeutic trial of levodopa should always be considered in SD patients. This type of dystonia is commonly diagnosed during childhood, between 4 and 8 years of age. Second-line treatment of paediatric dystonia is based on anticholinergic agents, the most common being benzhexol [8]. For severe dystonia episodes, Marsden et al. [3] suggest concurrent combined therapy with tetrabenazine, pimozide and benzhexol to achieve symptomatic relief. Some authors claim that more effective pharmacological therapies are based on baclofen, levodopa preparations and antiepileptics including carbamazepine [5].

Status dystonicus treatment should be conducted in the intensive care unit setting due to the risk of life-threatening complications and failure of attempted pharmacological therapies. The most serious immediate threat to the life of SD patients is respiratory failure triggered by the excessive contraction of muscles of the upper respiratory tract, chest and abdomen [1,7]. Status dystonicus patients are under the risk of developing dysphagia and aspiration caused by spasmodic action of the muscles of the throat and oesophagus which may lead to aspiration pneumonia [9]. Frequently, the symptoms listed above induce hypoventilation followed by hypoxaemia. This is also an immediately life-threatening condition requiring muscle paralysis, sedation, intubation and mechanical ventilation [1]. A major adverse complication is rhabdomyolysis (elevated levels of creatine kinase in blood serum) resulting from the breakdown or disintegration of striated muscles during their increased activity [1,2]. An early clinical sign of the condition may be discoloured (tea-coloured) urine. Rhabdomyolysis leads to sudden renal failure accompanied by anuria. In most instances, renal failure is transient with the eventual return of normal renal function; some patients, however, require temporary renal replacement therapy.

Exhaustion observed in SD patients as a result of prolonged severe dystonia and pain caused by dystonic spasms is recognized as an indication for performing sedation and muscle paralysis. Sedation and mechanical ventilation should be continued for 4–6 days. As the next stage, doses of sedatives and muscle relaxants are gradually reduced to assess the severity of SD. Effective control of SD symptoms over consecutive days can only be achieved with a sedative agent such as propofol. On account of its pharmacokinetic properties, the drug allows a quick assessment of the patient's consciousness and neurological status upon its discontinuation. Another drug routinely used for the sedation of SD patients is midazolam. As opposed to propofol, midazolam exhibits a slightly more pronounced sedative effect on the cardiovascular system. It also has a markedly longer half-life, which may occasionally interfere with the assessment of the patient's consciousness status [10,11].

3. Stereotactic ablative surgery (thalamotomy and pallidotomy) in the treatment of status dystonicus

In the past, stereotactic treatment was introduced in those SD patients who underwent sedation, muscle paralysis and mechanical ventilation, and experienced a recurrence of status dystonicus upon gradual discontinuation of sedatives and muscle relaxants. At the time, the stereotactic target of choice for the treatment of dystonia was the area of ventrolateral thalamic nuclei. Cooper [12] found thalamotomy to be the most effective method of treatment in patients with primary or secondary generalized dystonia. A 70% reduction of dystonia symptoms was achieved in the study conducted by this author. Cooper's excellent results, however, were not confirmed by findings obtained by Andrew et al. [13]. The authors noted only a 25% reduction of symptoms of primary or secondary generalized dystonia following thalamotomy [13]. Bilateral thalamotomy was associated with a very high risk of persistent postoperative dysarthria and dysphagia [13]. Thalamotomy was the first stereotactic surgery used in the treatment of patients with SD that was refractory to standard drug therapy [1]. In subsequent years, pallidotomy was found to be an effective modality in the treatment of drug-induced dyskinesia in patients with advanced Parkinson disease [14]. Consequently, the main target of stereotactic surgery shifted from the thalamus to the internal globus pallidus (GPi). The next stage in the development of functional neurosurgery for dystonia was the application of bilateral GPi stimulation for the treatment of primary generalized dystonia and then drug-refractory SD [15].

As mentioned previously, the first report of thalamotomy performed in two SD patients was published by Manji et al. [1]. Two-stage thalamotomy carried out in a 9-year-old boy with a history of dystonia and athetoid movements secondary to perinatal injury caused by premature birth failed to improve the patient's neurological status. A marked improvement following unilateral thalamotomy, however, was noted by the authors in a 38-year-old woman with right-sided post-encephalitic hemidystonia [1]. In addition to an improvement observed on the patient's right side, stereotactic left thalamotomy also improved the patient's dystonic position of the left upper limb. Another case of therapeutic benefit achieved with pallidotomy was reported by Justesen et al. [16] in a 10-year-old patient with NBIA and dystonia causing respiratory disability [16]. Unilateral (left-sided) pallidotomy brought about a regression of dystonic movements of the right side, and restoration of normal respiratory function. The effect proved to be long-lasting [16]. A similar benefit was achieved in the same indication by other authors following bilateral pallidotomy [17,18]. In a report of five cases of patients with SD episode, Teive et al. [18] describe bilateral pallidotomy in a patient suffering from cerebral palsy and generalized dystonia. The patient also had epilepsy. Bilateral pallidotomy combined with antiepileptic treatment, biperiden and clonazepam turned out to be an effective modality for controlling dystonic movements.

The cases of surgical treatment of SD outlined above support the efficacy of ablative operations (Table 1). The majority of patients underwent bilateral surgeries due to generalized severity of dystonic movements [16,18,19]. In some of the cases, unilateral stereotactic surgery was shown to be a therapeutic modality providing effective control of contralateral dystonic movements, contributing to a relief of SD symptoms [17,20]. Given limited experience, it is difficult to determine the preferred target for stereotactic surgery in SD treatment. Pallidotomy is more effective for SD management than thalamotomy, which stems principally from neuroanatomical considerations. Interestingly, the authors observed no complications of bilateral pallidotomies in the majority of children who underwent surgery due to primary generalized dystonia or SD [21]. Such complications – including dysarthria, dysphagia, sensory disorders and paresis – were reported in some patients following two-stage bilateral thalamotomy [13]. Furthermore, thalamotomy only involves the disruption of thalamocortical connections, without affecting pallidal fibres running from the main input structure for main output structure of the extrapyramidal system, i.e. the sensorimotor area of GPi which has a decisive effect on the pyramidal system [22]. In addition to blocking pallidal fibres which extend to the thalamus, and thalamocortical connections, pallidotomy also blocks pallidal and brainstem fibres running to the pedunculopontine nucleus which, as one of its primary functions, controls axial and proximal striated muscles via the descending reticulospinal tract. The muscles are very often involved in dystonic movements in patients suffering from different primary and secondary generalized dystonic syndromes. This explains the efficacy of pallidotomy or GPi stimulation in the treatment of dystonia involving axial muscles. A very interesting observation concerns bilateral reduction of hyperkinetic movements as a result of unilateral pallidotomy. The effect is markedly less common following unilateral thalamotomy [13,20]. It is determined by the presence of numerous pallidal fibres which extend not only to the ipsilateral thalamic nuclei including the complex of ventrolateral anterior and ventrolateral nuclei but also to the centromedian thalamic nucleus [21]. Neuroanatomical studies have demonstrated that 10–20% of all pallidal fibres transgress the median line in the posterior section of the centromedian thalamic nucleus and, via the supracommisural decussation, extend to the same thalamic nuclei as ipsilateral fibres [21]. In addition, pallidal and brainstem fibres innervate both ipsi- and contralateral pedunculopontine nuclei, i.e. bilateral motor regions of the brainstem. GPi, with its numerous connections to the thalamic nuclei and areas controlling brainstem motor function, is thus the stereotactic target of choice for the management of patients with dystonia and SD.

4. Deep brain stimulation in the treatment of status dystonicus

Similarly to ablative surgical methods, the number of SD patients treated with GPi stimulation continues to be limited. A review of international medical literature, however, reveals a growing number of reports on isolated cases or small groups of patients treated with GPi stimulation. Factors that have contributed to the wider use of GPi stimulation include increased availability of this neuromodulatory technique

Table 1 – Outcome of ablative procedures (thalamotomy, pallidotomy) in patients with status dystonicus. This table includes the information regarding aetiology of dystonia, coexistence of other involuntary movements, and triggering factors if identified. This table includes also data regarding first dystonic symptoms, age at surgery, results and follow-up time in each case described in the manuscript.

Authors and year of publication	Aetiology of dystonia and coexisting involuntary movements	Factor triggering status dystonicus	Age at the onset of dystonia [years]	Age at the time of stereotactic surgery [years]	Stereotactic target	Outcome of the surgery	Follow-up period (in months or years)
Manji et al. (1998) [1] Patient 1	Cerebral palsy followed by meningitis. Coexisting athetoid movements.	Probably a bacterial infection	2	13	Vim and Vop thalamic nuclei. Two-stage bilateral thalamotomy.	Poor	5 months – death of the patient
Manji et al. (1998) [1] Patient 2	Right-sided post-encephalitic hemidystonia	Probably a viral infection	17	38	Ventrolateral complex of thalamic nuclei – left-sided thalamotomy	Very good – bilateral effect	24 months
Justesen et al. (1999) [16]	Neurodegeneration with brain iron accumulation	The trigger of the SD episode has not been identified	6	10	Left-sided pallidotomy	Good	6 months
Kyriagis et al. (2004) [17]	Neurodegeneration with brain iron accumulation	The trigger of the SD episode has not been identified	Between 7 and 8	9	Bilateral pallidotomy	Good effect until 6 months of follow-up, then deterioration	15 months – death due to massive pneumonia
Teive et al. (2005) [18]	Dystonia secondary to cerebral palsy and epilepsy	The trigger of the SD episode has not been identified	No data available	8	Bilateral pallidotomy	Very good effect	Unknown
Balas et al. (2006) [19]	Neurodegeneration with brain iron accumulation	The trigger of the SD episode has not been identified	5	10	Ventrolateral complex of thalamic nuclei and ventrolateral posterior area of internal globus pallidus	Very good effect	48 months
Elkay et al. (2009) [28] Patient 1	Dystonia due to Batten's disease	The trigger of the SD episode has not been identified	5	17	Bilateral pallidotomy	Good	72 months

Vim – nucleus ventralis indermedius thalami; Vop –nucleus ventralis oralis posterior; SD – status dystonicus (dystonic state).

across the world, and its high efficacy in the treatment of primary or secondary generalized dystonias.

One of the first reports on successful SD treatment with bilateral GPi stimulation was presented by Angelini et al. [23] in a 13-year-old boy with a dystonia-dyskinesia syndrome. The aetiology of dystonia could not be determined. Most probably, the condition resulted from the patient's birth injury and his specific form of cerebral palsy. Bilateral GPi stimulation proved to be an effective method to control dystonic movements. Similar efficacy of bilateral GPi stimulation was reported by Zorzi et al. in three SD patients. The authors achieved an outstanding neurological improvement in two patients with DYT1-negative primary dystonia. In one patient with SD secondary to encephalopathy of unknown origin the improvement was, however, limited [24]. Teive et al. applied bilateral GPi stimulation to a 57-year-old female patient with cranial-cervical dystonia who had been treated with botulinum toxin injections for many years. The patient developed SD in reaction to a stress factor. Bilateral GPi stimulation brought about a total regression of dystonia [18].

The efficacy of GPi stimulation for the management of SD secondary to NBIA was confirmed by reports authored by Mariotii et al. [10] and Grandas et al. [25]. Very high efficacy of bilateral GPi stimulation in the treatment of primary generalized dystonia without DYT-1 mutation was also reported by Jech et al. [26]. Bilateral implantation of electrodes in the internal globus pallidus in a 12-year-old boy resulted in a complete regression of SD [26]. One of the factors potentially triggering SD is an abrupt change in drug therapy, while dystonia can be a consequence of prolonged treatment with antipsychotic agents, particularly antagonists of dopamine receptors. The drugs, which are usually taken on a long-term basis, may lead to a range of motor disorders including dystonia, Parkinson disease and akathisia. SD induced by antipsychotic drugs is a rare phenomenon. One such case was reported by Kovacs et al. [27]. Prolonged use of risperidone combined with clonazepam led to segmental dystonia which escalated into SD. Bilateral GPi stimulation contributed to a total regression of dystonia.

A very interesting report on the application of a combined neurosurgical modality comprising pallidotomy and bilateral GPi stimulation for the treatment of SD due to Batten's disease was presented by Elkay et al. [28]. The authors argue that combined neurosurgical treatment remains an effective method of treatment of hyperkinetic movements leading to SD in paediatric patients with Batten's disease. It improved the quality of life of patients and allowed their return to normal life. What is more, pallidotomy or GPi stimulation makes it possible to gradually discontinue sedative drugs, and contributes to the subsidence of painful muscle spasms and symptoms of generalized dystonia [28].

A report on the cases of two patients with dystonic cerebral palsy and coexisting choreoathetosis, who developed status dystonicus after undergoing general anaesthesia, was published by Apetauerova et al. [29]. A follow-up period of several months demonstrated that bilateral GPi stimulation had produced an immediate resolution of SD in both cases. In the latest report, dating from 2012, Walcott et al. [30] discuss cases of three children with status dystonicus resulting from various dystonic syndromes that were treated with bilateral GPi stimulation. The technique proved to be the most effective method of SD treatment. All studies investigating GPi stimulation in SD treatment are listed in chronological order in Table 2.

One of the most important aspects that must be addressed when considering implantation of a deep brain stimulation system in children is their continued growth. Steady brain growth in paediatric patients may cause dislocation of the electrode, which is associated with an elevated risk of a gradual loss of stimulation efficacy. None of the studies outlined above mentions the problem, probably because the majority of surgically treated SD patients were over 12 years old and only 4 patients underwent an operation before 10 years of age. The youngest surgically treated patient was 8 years old [24,30]. Furthermore, postoperative follow-up period among the youngest patients treated with bilateral brain stimulation varied from 3 months to 19 months. Thus, it was impossible for the intracranial electrode to become dislocated as a result of brain growth over such a short follow-up period [24,30]. The potential problem can be resolved by changing active contacts of the electrode implanted in the brain. When GPi is the stereotactic target, the distance between the active contacts of the electrode is 1.5 mm. By stimulating proximal contacts, it is possible to compensate for possible dislocation of the electrode in the brain caused by children's growth. If, however, the effect of stimulation and the possibility of managing dystonia by changing active contacts become significantly compromised, the solution to consider is the implantation of another electrode. The method was applied by French neurosurgeons in patients with generalized dystonias in whom the therapeutic effect of GPi neurostimulation had become reduced. The implantation of another electrode caused further improvement of the patients' clinical condition [31]. Another possible approach is to move the initially implanted electrode upwards in the stereotactic setting under fluoroscopic control. The technique, however, is associated with an increased risk of intracranial bleeding. Children's physiological growth is also a factor potentially leading to a higher risk of mechanical damage to the deep brain stimulation system in children. Currently available deep brain stimulation systems are more flexible and elastic, which, however, does not completely rule out the possibility of mechanical disruption caused by brain growth. American authors who applied the method of brain stimulation in children described the complication as quite rare: it was noted in just two cases (one case of intracranial electrode break and one of damage to the extension) in the initial study group of 31 children. The mean age of the patients at the time of surgery was 13.2 years, and varied from 4 to 17 years. The most common complications observed by the authors were infections, especially in the youngest patients. In order to reduce the risk of infection, mechanical damage or skin erosions developing above the implant, the majority of procedures in children involve placement of the pulse generator in the abdomen [30,32]. Implantation of the pulse generator subcutaneously below the clavicle is not recommended because the device is relatively large which, coupled with the thinness of the subcutaneous fatty tissue, makes the site prone to erosions and infections [24,30].

Table 2 – Outcome of deep brain stimulation – bilateral pallidal stimulation in patients in dystonic state. The information included in this table describes the same data like in Table 1.

Authors and year of publication	Aetiology of dystonia and coexisting involuntary movements	Factor triggering status dystonicus	Age at the onset of dystonia [years]	Age at the time of stereotactic surgery [years]	Stereotactic target	Outcome of the surgery	Follow-up period (in months or years)
Angelini et al. (2000) [23]	Cerebral palsy	The trigger of the SD episode has not been identified	8 months	13	Bilateral GPi stimulation	Very good	7 months
Zorzi et al. (2005) [24] Patient 1	Primary generalized non-DYT-1 dystonia	The trigger of the SD episode has not been identified	1.6	8.2	Bilateral GPi stimulation	Very good	15 months
Patient 2	Encephalopathy of unknown origin	Upper respiratory tract infection	6 months	14.2	Bilateral GPi stimulation	Slight	15 months
Patient 3	Primary generalized non-DYT-1 dystonia	The trigger of the SD episode has not been identified	3	10.6	Bilateral GPi stimulation	Very good	19 months
Teive et al. (2005) [18]	Craniocervical dystonia	Stress factor	Long history of dystonia	57	Bilateral GPi stimulation	Very good effect	Unknown
Mariotii et al. (2007) [10]	Neurodegeneration with brain iron accumulation (PANK2)	Upper respiratory and urinary tract infections	7	15	Bilateral GPi stimulation	Good	12 months
Elkay et al. (2009) [28] Patient 2	Dystonia due to Batten's disease	The trigger of the SD episode has not been identified	2	19	Bilateral pallidotomy + bilateral GPi stimulation	Good	7 months
Jech et al. (2009) [26]	Primary generalized dystonia	The trigger of the SD episode has not been identified	8	12	Bilateral GPi stimulation	Good	15 months
Apeetauerova et al. (2010) [29] Patient 1	Cerebral palsy	General anaesthesia + metoclopramide	16	16	Bilateral GPi stimulation	Very good	30 months
Patient 2	Cerebral palsy	General anaesthesia	26	26	Bilateral GPi stimulation	Very good	34 months
Grandas et al. (2011) [25]	Neurodegeneration with brain iron accumulation (PANK2)	The trigger of the SD episode has not been identified	17	19	Bilateral GPi stimulation	Very good	9 months
Kovacs et al. (2011) [27]	Drug-induced dystonia	Antipsychotic drugs	18	18	Bilateral GPi stimulation	Very good	12 months
Walcott et al. (2012) [30] Patient 1	Neonatal jaundice, encephalopathy, cerebral palsy	Spinal surgery followed by infection of surgical wound	Long history	14	Bilateral GPi stimulation	Very good	12 months
Patient 2	Primary generalized dystonia	The trigger of the SD episode has not been identified.	4 months' history	9	Unilateral GPi stimulation	Good	2 months
Patient 3	Cerebral palsy, spastic quadriparesis	Probably an upper respiratory tract infection	Exacerbation of dystonia over preceding two months	9	Bilateral GPi stimulation	Good	3 months

SD – status dystonicus (dystonic state); GPi – internal globus pallidus.

Polish reports on deep brain stimulation fail to include any mention of the treatment of SD in any patient suffering from generalized dystonia. Polish publications on GPi stimulation in the management of dystonia confirm that the method has been both effective and safe for the treatment of diverse motor disorders including advanced Parkinson disease over the past several years [33–35].

5. Summary

The cases of surgical treatment of SD presented above show that ablative operations and GPi stimulation are effective therapeutic modalities. Status dystonicus always requires combined treatment, whereas stereotactic surgery is regarded as the method of last choice for SD. Neurosurgical treatment is beneficial in that it eliminates the need for using sedative drugs and successfully alleviates symptoms accompanying primary or secondary generalized dystonias. Nearly 60% of SD cases occur in children below 15 years of age. The therapeutic method of choice remains bilateral GPi stimulation. What is more, the method can be successfully applied in patients who have previously had ablative surgery (either thalamotomy or pallidotomy), as shown in medical reports. Neurosurgery is not only a highly effective form of SD treatment but also prevention: no SD recurrence has been observed in patients with a history of neurosurgical procedure.

Conflict of interest

None declared.

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Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

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