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Surgical management of hip instabilities in children with spina bifida

Spina bifidalı çocuklarda kalça instabilitesinin tedavisi

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Amaç: Kalça instabilitesi nedeniyle ameliyat edilen spina bifidalı çocuklarda cerrahi tedavi sonuçları değerlendirildi.

Çalışma planı: Kalça instabilitesi (subluksasyon/çıkık) nedeniyle 26 hastanın (16 kız, 10 erkek; ort. yaş 4.5; dağılım 3-6) 28 kalçası ameliyat edildi. Yirmi dört çocukta alçak seviye (L_4-sakral) lezyon ve yürüme potansiyeli (2 olguda iki taraflı çıkık) vardı; bunların 16'sında tek taraflı çıkık yanı sıra bacak boyu eşitsizliği (>2 cm) ve kompansatuvar skolyoz görüldü. Yüksek seviye (torasik-L_3) tutulum ve tek taraflı çıkık olan iki çocukta yürüme yoktu. Yüksek seviye tutulumu olan iki hastada ve alçak seviye lezyonlu hastaların bir kısmında (9/26 kalça) kalça fleksiyon kontraktürü de saptandı. Kalça stabilitesinin sağlanmasına yönelik olarak açık redüksiyon, pelvik osteotomi, gerektiğinde femoral osteotomi ve pelvipedal alçı uygulamasına başvuruldu. Hastalar ortalama 38 ay (dağılım 30-48 ay) süreyle klinik ve radyografik olarak izlendi.

Sonuçlar: Bacak boyu eşitsizliği ve skolyoz olan 16 hastanın 14'ünde yürüme fonksiyonlarında gelişme görüldü; iki hastada eşitsizlik ve skolyoz devam etti. Diğer hastalarda, ameliyat öncesi fonksiyonel durumların ameliyat sonrasında korunduğu görüldü. Ameliyattan sonra kalçaların ortalama hareket açıklığı azaldı, fakat hiçbirinde eklem sertliği gelişmedi. Üç olguda yüzeyel yara enfeksiyonu, iki olguda distal femoral diyafiz kırığı dışında ameliyat sonrası erken dönemde komplikasyon gözlenmedi. Üç hastanın implantları, cilt altında belirginleşmeleri nedeniyle altıncı ayda çıkartıldı. Üç hastanın geç dönem radyografilerinde, girişim gerektirmeyen subluksasyon tekrarı gözlendi.

Çıkarımlar: Spina bifidalı çocuklarda, yüksek seviye lezyonlara bağlı kalça sorunlarında cerrahi tedaviden genellikle kaçınmak gerekirken, alçak seviye lezyonlarla birlikte olan kalça instabilitesinde başarılı sonuçlar alınabilmektedir.

Anahtar sözcükler: Çocuk; kalça çıkığı/etyoloji/komplikasyon/cerrahi; kalça eklemi/fizyoloji; bacak uzunluğu eşitsizliği/etyoloji; omurga disrafizmi/komplikasyon/cerrahi; tedavi sonucu.

Objectives: We evaluated the results of surgical management of hip instability in children with spina bifida (SB).

Methods: Twenty-eight hips of 26 patients (16 girls, 10 boys; mean age 4.5 years; range 3 to 6 years) were surgically managed for hip instability (subluxation/dislocation) associated with SB. Twenty-four patients (2 bilateral dislocations) had low-level lesions (L_4-sacral) and a potential to walk, of which 16 patients presented with unilateral dislocation with functional problems including significant (>2 cm) limb-length discrepancy and scoliosis. Two patients had high-level lesions (thoracic-L_3) associated with unilateral dislocations and were unable to walk. Those with a high-level lesion and some patients (9/26 hips) with a low-level lesion also had hip flexion contractures. Treatment included open reduction, pelvic osteotomy, proximal femoral osteotomy when necessary, and a spica cast. The patients were clinically and radiographically monitored for a mean of 38 months (range 30 to 48 months).

Results: Of 16 patients with functional problems, 14 patients had improvement in their gait patterns, while limb-length discrepancy and scoliosis persisted in two. The remaining 10 patients maintained their preoperative functional statuses. The mean range of motion of the hips decreased postoperatively; however, none of them developed joint stiffness. Early postoperative complications included superficial wound infections in three patients, and distal femoral diaphyseal fractures in two patients. Three patients required removal of the implants in the sixth month due to subcutaneous prominence thereof. Late radiographs of three patients showed recurrent subluxations, which did not require any intervention.

Conclusion: Although surgical treatment of hip problems associated with high-level lesions may be unrewarding in children with SB, those associated with low-level lesions can be successfully managed with proper surgical indications.

Key words: Child; hip dislocation/etiology/complications/surgery; hip joint/physiology; leg length inequality/etiology; spinal dysraphism/complications/surgery; treatment outcome.

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Spina bifida (SB) is a condition leading to disorders in the neuromuscular function due to the incomplete development of any area in the spinal cord. Children with spina bifida experience various orthopaedic problems throughout their life; some major ones include hip deformities and hip instability. 30-50% of these children with spina bifida develop hip subluxation or dislocation during the first two and three years of their life.[1,2] Those hip problems result from the weakness of the muscles surrounding the hip, imbalanced power among the muscle groups and other associated structural problems.[1]

The orthopaedic problems of the children with spina bifida should be considered as a whole, and it should be kept in mind that the problems related with the hip are only a part of that whole. The benefit/harm ratio of the treatment should be well calculated, particularly if a surgical treatment is planned. Literature analysis showed that the most important criteria defining the treatment for hip instability in children with SB are the level of neurologic involment, child's potential to walk, and unilaterality or bilaterality of the instability. Surgical interventions usually produce successful results in patients with low- level lesion (lower lumbar, sacral) and potential to walk while it is frequently unsuccessful in patients with no potential to walk associated with high-level lesions (toracal, upper lumbar).[1-6]

In this study, we evaluated mainly the results of the surgical treatment in a limited number of patients who had L_4-sacral neurologic involvement, and were ambulatory and had unilateral hip instability, and also the results of the surgical treatment in a limited number of patients with partial surgical indication (bilateral involvement associated with L_4-sacral lesion or unilateral involvement associated with toracal- L_3 lesion).

Patients and method

Twentyeight hips of 26 patients (16 girls, 10 boys; mean age 4.5 years; range from 3 to 6 years) who presented to the Multidisciplinary Spina Bifida Clinic between 1998 and 2001 for hip instability (subluxation/dislocation) were operated. Clinical and radiographic data were obtained, all patients undergoing preoperative evaluations and regular postoperative follow-ups. The patient group consisted of children who previously underwent various surgical procedures other than orthopaedic. The mean number of those operations, mainly urologic and neurosurgical, per patient was two (1 to 4 operations). The sac-closure operation of all patients was performed during the first days of the delivery. Some of the children were treated with several conservative methods for hip deformities such as physical treatment.

The level of neurologic involvement, potential to walk and distribution of the hip instability (unilateral or bilateral) were recorded. The level of neurologic involvement was separately determined for both sides. Based on this, most of the patients (24 patients; 14 girls/10 boys) had low-level (L_4-sacral) lesion. The hip instability was unilateral in 22 of the patients with potential to walk, and bilateral in two (Figure 1a, 3a). Majority of the children with low-level involvement used AFO (ankle-foot orthosis). High-level neurologic involvement (toracal-L_3) was seen in two patients. The hip instability was unilateral in those patients with no potential to walk in the long period (Figure 2). In addition to the hip instability, hip flexion contracture was also found in two patients with high-level involvement and some of the patients with low-level lesions (9/26 hips). Presence of hip flexion contracture was the main indication for surgical treatment in some of those patients.

Motion range of the hips, limb-length discrepancy (BBUF), motion range of the knees, presence of scoliosis and pelvic obliquity and muscle strengths were determined by physical examination. Those assessments were recorded both during the preoperative examination and postoperative follow-up period. The neurologic status of the motor involvement was evaluated testing the muscle strength by hand. It was observed that most of the patients with unilateral instability (16/24) had functional problems. They had clinically remarkable (>2 cm) limb-length discrepancy (usually evident in dislocated hips) in varying degrees and associated pelvic obliquity and nonstructural compensatory scoliosis. No functional problem was seen in a few patients with bilateral instability (2 patients). During the postoperative follow-up period, it was investigated if those pathologies continued. Motion ranges for hips and knees

were controlled, and changes in the montion range of the joints after the operation were recorded.

The hip graphs were evaluated during the pre- and post-operative follow-up period, and dislocated, subluxed or normal hips were recorded. Based on this, 18 hips were found dislocated and 10 hips were found subluxated during the preoperative radiographic evaluations. Postoperative radiographic follow-ups were performed immediately after the operations, and then at months 4.5, 3, 6 and 12 and yearly.

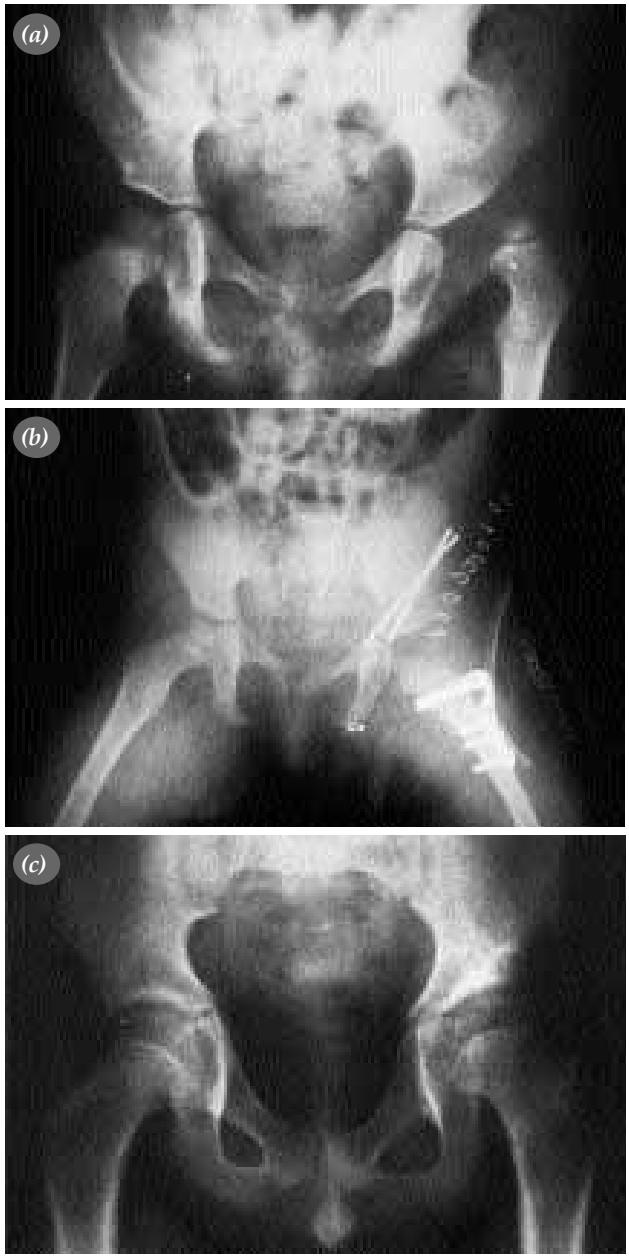

Figure 1. A four-years old boy with a unilateral hip insta bility associated with low-level lesion. **(a)** The dislocation in the left hip is evident in the antero-posterior pelvic view. **(b)** The patient underwent open reduction, pelvic osteotomy (Salter osteotomy), proximal femoral varus derotation osteotomy,followed by a pelvipedal cast. **(c)** A view taken three years later showed that concentric reduction of the operated hip is well maintained.

Table 1. Surgical procedures implemented on hips

	Low-level lesion (L_4-sacral)	High-level lesion (toracal-L_3)
Open reduction	26	2
Pelvic osteotomy	26	2
Proximal femoral osteotomy	15	1
Release for flexion contracture	9	2

For hip stability, open reduction, pelvic osteotomy, femoral osteotomy and pelvipedal cast, when required, were performed (Table 1). Detailed information related with those operations was obtained from the patient files. Pelvic osteotomies included acetabuloplasties (Salter, Pemberton and Pembersal) while varus derotation osteotomy was performed when proximal femoral osteotomy was needed (Figure 1b, 3b). Accordingly, acetabuloplasty was performed in all of 28 hips in addition to the open reduction; Salter osteotomy was performed in most of the cases (24/28) while Pemberton (2/28) and Pembersal (2/28) osteotomies in others. Proximal femoral varus derotation osteotomy was required in

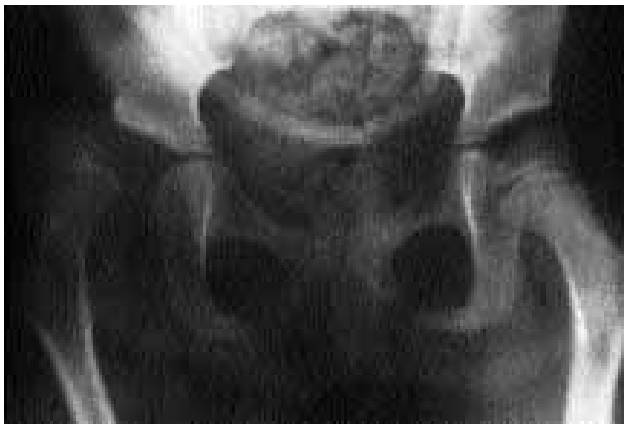

Figure 2. A four-years old boy with a unilateral hip insta bility associated with high-level lesion. The dis location in the right hip is evident in the antero-posterior pelvic view.

more than half of the cases (16/28). Femoral reduction was performed in a very few operations (3/28). Soft tissue release was performed in patients with remarkable hip flexion contracture; no osteotomy was needed. As implant, K-wire was used in pelvic osteotomies, when required, while Harris plaque (pediatric size) was employed in proximal femoral osteotomies. Pelvipedal cast was used following all of the operations. The cast was changed at postoperative month one and a half under general anaesthesia as to reach three months at total, which was followed by a device for a period of three months. Cast care was instructed to the parents in detail before discharging the patient, and cleanliness of the cast was controlled at all follow-ups. Some of the patients were included in a short-term rehabilitation program to enhance the motion range of the joints (since there was no joint stiffness, only decrease in the motion range). However, in general, motion range exercises at home supervised by parents were efficient. The hips of patients with bilateral involvement were operated in separate sessions; operations were carried out with a mean interval of 10 months (range from 8 to12 months).

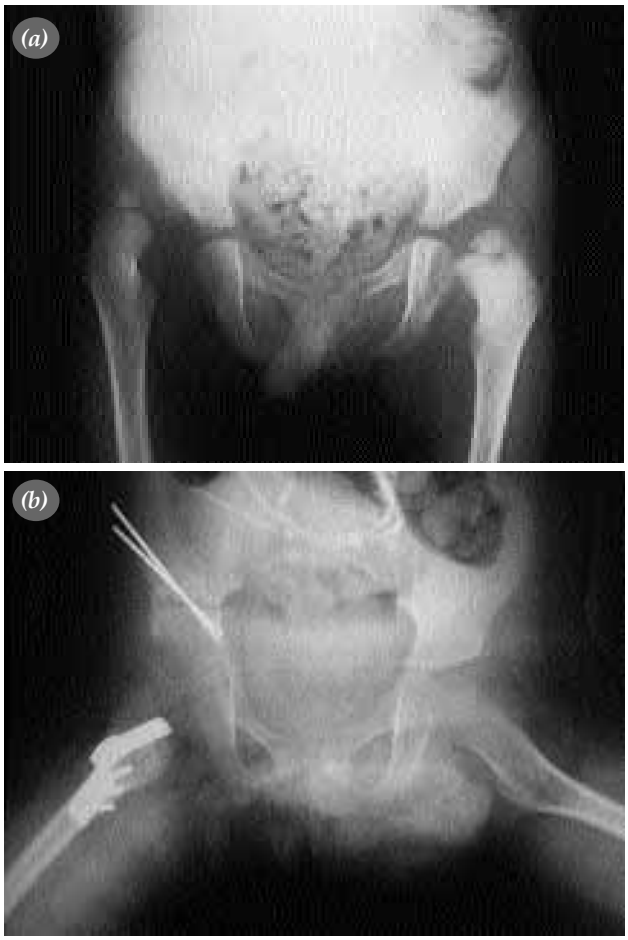

Figure 3. A three-years old girl with bilateral hip instability associated with low-level involvement. **(a)** Subluxation in the left hip and dislocation in the right hip are evident in the antero-posterior pelvic view. **(b)** The right hip of the patient under went open reduction, pelvic osteotomy (Salter osteotomy), proximal femoral varus derotation osteotomy followed by pelvipedal cast. Her left hip was operated with similar procedures eight months after this operation.

The patients were followed clinically and radiographically (Figure 1c). Postoperative early and late complications were recorded. Presence of complications such as development of deep superficial infection or hematoma, reduction loss and implant (fixation) failure was investigated during the early postoperative period while during the late period, presence of infection again, problems associated with the implant and reduction loss in the radiographic follow-ups were examined.

Furthermore, treatments implemented for developing complications were recorded. The mean follow-up period was 38 months (range from 30 to 48 months).

Results

Fourteen of 16 children who had unilateral hip instability associated with low-level involvement and functional problems had improvement in the gait pattern during the postoperative period. It was observed that the limb-length discrepancy was removed to a large extent or came to a point where there was no clinical difference (<2 cm) after the hip reduction in those children, who usually had dislocation in the hip. Removal of the limb-length discrepancy also resulted in improvement of the problems like pelvic obliquity and nonstructural scoliosis, which, in turn, enabled the patients to ambulate more comfortably, to get tired less often and to spend less energy. No change was observed in the functional status of other two patients: Limb-length discrepancy and nonstructural scoliosis continued after the operations. All remaining patients (10/26) maintained their existing functional status during the postoperative period. The mean motion range of the operated hips was reduced in all directions during the postoperative

follow-ups, however no joint stiffness developed in any of the patients.

None of the patients had reduction loss or implant failure (fixation failure) at the early postoperative period. No serious wound problems like deep infections or hematoma were seen; three patients developed superficial wound infection; those conditions responded pretty well to the treatment by oral antibiotics, and improved within 1-2 weeks. Plain graphies taken upon detecting a swelling and rash at the femoral distal during the exchange of pelvipedal cast in one of the children one and a half month after the operation demonstrated a united (together with a giant callus formation) fracture. This fracture, which was not evident in the preoperative views, was considered to occur during the operation (probably during the build-up of the pelvipedal cast). No change occurred at the prognosis of the child since the fracture was united during the implemetation of the pelvipedal cast. Another patient developed a mild deplaced diaphyseal fracture again at the femoral distal during the rehabilitation at the postoperative month 4. Long-limb splint was used for this child, and he was returned to the rehabilitation after the union of the fracture. No joint stiffness developed in this patient; motion range of the hip joints was reduced like other patients. Therefore, it was proposed that development of the fracture didn't affect the prognosis in this child very much.

The implants of three patients were removed after the union was demonstrated by the radiographies at the postoperative month 6 since they had become subcutaneously obvious. Following the implant removal operations, no complication was seen at the early or late period. Radiographically it was observed that subluxation was recurrent in three patients at the late period (within 1st year in one patient, and within 2nd year in two patients). They were not intervened, considering that it wouldn't make any change in the functional status of the children. No progress was found in the clinical and radiographical follow-ups of those children.

Discussion

Spina bifida means maldevelopment of any area in the spinal cord. It is also called neural tube defect, myelomeningocele and spinal dysraphism. It rates second among the diseases leading to disorders in the neuromuscular functions in children after serebral palsy. The main problem is the failure of closure occured in the midline while the antecedent structure called neural placode is forming a tube during the formation process of the spinal cord. As a result of this failure, a bone protrudes behind the canal of the vertebrae. The most evident finding in spina bifida is the motor dysfunction, expressed as paraplegia or paraparesia. Motor dysfunction can be seen immediately after the delivery depending on the level of effect on the spinal cord and meninges as well as occurring in advanced ages due to tension in the cord.[1,2]

Various orthopaedic problems are experienced in the children with spina bifida depending on the degree of the neurologic involvement. The leading ones include deformities of the vertebrae and lower extremities and joint contractures. Orthopaedic problems have remarkable effect on the quality of child's life and the prognosis. The objectives of the orthopaedic treatment are to achieve the upper potential mobility level and to prevent a series of complications that this condition may lead to.

30-50% of children with spina bifida develop hip problems, hip instability (subluxation/dislocation) and flexion contracture during the first two-three years of their life.[1,2] Hip problems are rare with sacral lesions while it reachs up to 70% in upper lumbar and toracal lesions.[1,7] Before, it was thought that only weakness of some muscle groups surrounding the joint and associated imbalance were responsible from the hip problems experienced in those children. It was believed that the affecting hip flexors (with or wihout active adductors) caused the instability alone in the absence of extensors and abductors. However, as a result of follow-up of the patients with SB for long years, it has been observed that the incidence of developing instability in patients with evident imbalanced muscles is not as high as expected, and children with no muscle imbalance frequently developed dislocations. Therefore, today it is believed that muscle imbalance is not the only factor, even tough important, in the development of instability.[1] The structural changes occurring along with

the muscle imbalance contribute to the development of hip problems. Major structural changes include coxa valga, capsule laxicity, increased femoral anteversion and defected acetabular development.[8]

In the past, it was believed that children with SB must walk for the sake of their general status, and no hip problems like instability or deformity must have been present for a proper gait pattern. Therefore, reduction of the hip dislocation was suggested in order to improve the functions of these children and to prevent the complications associated with hip dislocations. However, in the last two decades the validity of this approach has been intensively questioned, and it was observed that the functions became worser in some children with SB following the hip surgery. Furthermore, it has been revealed that the hip dislocation has a very little effect on the gait pattern.[1,9] It was observed that the walking difficulties in such patients mainly result from the permanent flexion deformity in the hip and associated excessive lumbar lordosis. Another significant point is the conclusion that it is not necessary for all children with SB to walk in the long term. Patients with high-level SB cannot usually walk after ten years old; mobilization is easier with wheel chair for most of those children and energy expenditure is more economic.[1,8]

Based on this information, in recent years some important issues were set in defining the surgical indications for hip instability in children. Major issues include neurologic level of the motor involvement, gait potential of the patient, extension of the involvement, and unilaterality or bilaterality of the hip dislocation.[1-3] The motor involvement of the muscles around the hip increases as the level of the lesion rises, which, in turn, has a negative effect on the gait potential of the child. Particularly the quadriceps muscle group, very important for the walking function, has been affected more in high-level lesions and ambulation becomes impossible in the long run. Patient's expectation for ambulation is an important factor in defining the surgical indication; reduction for hip dislocation is not suggested for patients with no potential to walk while stabilization of the instable hips improves the functional status of the walking patient.

Unilateral hip dislocation generates limb length discrepancy in children with SB in varying degrees. Furthermore, it may lead to pelvic obliquity, nonstructural scoliosis and iscial ulcers (associated with sitting long hours in the presence of hip asymmetry) in some of the patients. When unilateral hip dislocation is combined with low-level lesions, the objective of the treatment for hip is to improve the functional capacity of the child and prevent the development of those potential problems. Hip reduction usually provides successful results in such cases.[1-3,6] In case of unilateral hip dislocation associated with a high-level lesion, functional problems are rare, and surgical interventions frequently fail. Unilateral hip dislocation combined with high-level lesions has surgical indication since those children has no possibility to walk in the long term; those dislocations, for example, when hip flexion contracture would be surgically treated, are reduced.[1,3,10,11]

In this study, we evaluated the children with SB and hip problems, who had surgical treatment indications in accordance with the general principles defined in the literature. Majority of the patients undergoing surgical treatment consisted of the children with unilateral hip instability associated with low-level lesion. Surgical treatment was successful in most of these patients, and functional status was enhanced as defined in the literature. However, it was observed that some functional problems like limb length discrepancy and scoliosis continued in some of the patients. There is partial indication for surgical_treatment in bilateral dislocations associated with low-level lesions and unilateral instabilities accompanying high-level involvement. In our study, surgical stabilization was used since the operation of few such patients was planned for flexion contracture. The functional status of those patients didn't change, but stabilization of the hips was maintained during the follow-up.

The aim of the treatment in spina bifida is to make the child a comfortable, painfree, mobile and sociable individual. The approach for hip problems in those children has undergone a significant change in the recent years along with better com-

prehension of the formation mechanisms and results of long-term follow-ups. The indications for hip surgery in children with spina bifida are defined better so that improved results are obtained.

Surgical treatment should be avoided in hip problems developing as a result of high-level lesions while successful results are achievable in hip instability combined with low-level lesions.

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