Case Report

International Journal of Spine Surgery and Research

The Results of Vertebral Column Decancellation (VCD) For Correction of Thoracolumbar Kyphosis in an Achondroplastic Patient with Severe Hypoplastic Vertebra

Mirzashahi Babak[1], Malekzadeh Maziar[1]* and Saied Besharaty[2]

[1]*Associated Professor, Department of Orthopaedic Surgery, Tehran University Medical Science, Iran.*

[2]*Department of Orthopaedic Surgery, Atieh Hospital, Tehran, Iran.*

***Correspondence:**
Malekzadeh Maziar, Department of Orthopaedic Surgery, Tehran University of medical sciences, Iran.

Received: 25 Jun 2024; **Accepted:** 15 Aug 2024; **Published:** 22 Aug 2024

Citation: Babak M, Maziar M, Besharaty S. The Results of Vertebral Column Decancellation (VCD) For Correction of Thoracolumbar Kyphosis in an Achondroplastic Patient with Severe Hypoplastic Vertebra. Int J Spine Surg Res. 2024; 1(1): 1-5.

ABSTRACT

Background: *Achondroplasia is the most common cause of skeletal dysplasia and thoracolumbar kyphosis is one of the most common spinal disorders in achondroplastic patients. If the thoracolumbar kyphosis progresses despite conservative treatment, or if the kyphosis exceeds 50 degrees, or patients develop neurological deficits relating to the thoracolumbar kyphosis, surgical intervention is indicated. Currently there are several surgical techniques for correction of thoracolumbar kyphosis, such as pedicle subtraction osteotomy (PSO) and vertebral column resection (VCR) but there are few reports about using Vertebral Column Decancellation (VCD), especially in skeletally mature achondroplastic patients. The purpose of this study is to evaluate the results and complications and outcome of Vertebral Column Decancellation (VCD) in a skeletally mature achondroplastic patient.*

Case Presentation: *A 34 years old achondroplastic man with severe thoracolumbar kyphosis (103 degree) and significant neurological symptoms such as bilateral lower limbs numbness and claudication underwent Vertebral Column Decancellation (VCD). The patient was evaluated radiographically and clinically for 2 years with SRS-22 Score and ODI Score and VAS Score. The patient responded very well to VCD. The neurological symptoms improved and deformity was corrected with minimal surgical complications.*

Conclusions: *The long term radiological and clinical results of Vertebral Column Decancellation (VCD) in treatment of sharp angular thoracolumbar kyphosis in achondroplastic mature patient demonstrated that this technique is a relatively safe procedure and also has favorable outcomes. Since surgical complications are the major concerns about 3-column osteotomies, strict adherence to agreed surgical protocols can minimize the rates of complications (such as dural rupture, surgical site infection, massive bleeding). With meticulous hemostasis and administration of tranexamic acid during the surgery the rate of blood loss can be reduced. Despite the impaired enchondral ossification in achondroplastic patients, due to the bone-to-bone contact in VCD, the risks of nonunion and subsequent complication such as rod breakage can be reduced.*

Keywords

Achondroplasia, Thoracolumbar kyphosis, Vertebral Column Decancellation.

List of Abbreviations

VCD: Vertebral Column Decancellation, SVA: sagittal vertical axis, TLSO: Thoracolumbosacral, PSO: Pedicle subtraction osteotomy, VCR: VertebraL column resection, SPO: Smith-Petersen osteotomy.

Introduction

Achondroplasia is one of the most common cause of skeletal dysplasia [1-5]. Achondroplasia is inherited in an autosomal dominant trait and caused by a mutation of the FGFR3 [6,7].

This mutation results in failure of chondrocyte differentiation and endochondral ossification [8]. Clinical manifestations of achondroplasia such as rhizomelic short stature and frontal bossing, depressed nasal bridge, and hands with 3-pronged (trident) appearance are due to impaired cartilage production and enchondral ossification [3,9].Spinal canal stenosis and thoracolumbar kyphosis are the most common abnormalities affecting the achondroplastic spine. Spinal canal stenosis is due to shortened pedicles and decreased interpedicular distances [2]. Thoracolumbar kyphosis is a developmental deformity and the kyphosis is non-congenital and is not associated with structural defect of the vertebra [8]. The thoracolumbar kyphosis is present at birth and usually progresses at the age of 6-18 months and kyphosis is most evident when the patients start to sit up. Usually with improvement of muscle tones after standing and walking the deformity resolves spontaneously [3,8]. If the kyphosis progresses and became greater than 30 degrees TLSO bracing is recommended [10]. Surgical intervention is required for the patients with progressive kyphotic deformity and for those who develop significant neurological deficits [2]. Currently several surgical techniques such as Pedicle subtraction osteotomy (PSO) and vertebral column resection (VCR) are used for correction of thoracolumbar kyphosis [8,11-13] but there are few reports about using Vertebral Column Decancellation (VCD) technique for treatment of achondroplastic patients, therefore the purpose of this study is to evaluate the advantage and disadvantages of this procedure on a 34 years old achondroplastic patient with severe thoracolumbar kyphosis.

Case Presentation

The patient is 34 years old achondroplastic man with severe thoracolumbar kyphosis (103 degree) and significant neurological symptoms such as lower limbs numbness and claudication, without urinary or fecal incontinence and without motor weakness was referred to our institute. The patient's height was 128 cm. He had characteristic achondroplastic facial features such as prominent forehead and depressed nasal bridge. His arms and legs were short he had trident hands. Deep tendon reflexes were normal on both sides and babinski reflex was negative bilaterally. The muscle power was 5 for both legs. Severe thoracolumbar kyphosis and lumbar hyperlordosis were evident (Figure 1).

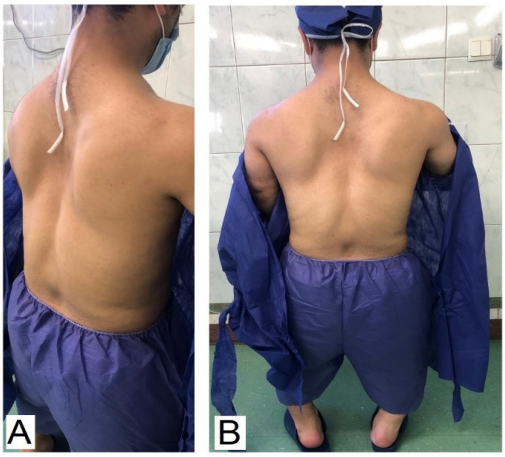

Figure 1: Preoperative photos of the patient.

Preoperative radiography showed severe thoracolumbar kyphosis; and T12-L2 segmental kyphotic angle was 103 (figure 2). Preoperative lumbar lordosis (L1-S1) was 81.5 and thoracic kyphosis (T1-T12) 18.6 and (T5-T12) regional kyphosis was 17.5 and SVA was -45mm. Preoperative MRI showed severe stenosis at (T7-L5) levels and CT scan showed hypoplastic wedge shape T12 and L1 vertebra.

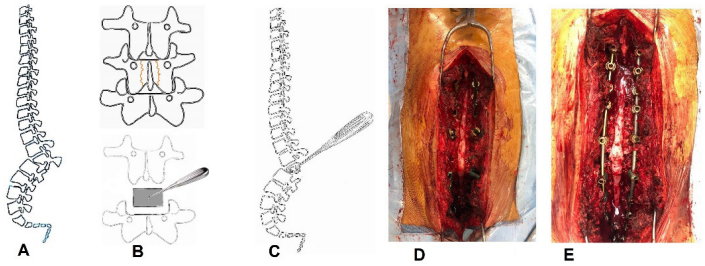

Figure 2: A-B Preoperative radiography and CT scan of the patient show a sever hypoplastic vertebra and thoracolumbar kyphosis. C-D sagittal MRI shows severe spinal canal stenosis in thoracolumbar and lumbar areas.

After performing necessary preoperative clinical and evaluation, informed consent was obtained from the patient and the surgery was performed in Atieh Hospital in Tehran, Iran in February 2020.The operation was performed under neuromonitoring [somatosensory-evoked potentials (SEPs) and motor-evoked potentials (MEPs) during the operation] [14]. In order to reducing the blood loss during the surgery Tranexamic acid [15] was administered parenterally and meticulous hemostasis was performed. After general anesthesia, the patient was placed prone on the radiolucent table. A standard posterior middle incision was made and by subperiosteal dissection of the paravertebral muscles the posterior elements of spinal column from T7 to S1 level was exposed then the Pedicle screws were placed from T7 to S1 levels by the freehand technique and using C-arm fluoroscopy to confirm the proper insertions. Due to severe spinal canal stenosis in thoracic and lumbar area decompressive laminectomy [16] were performed from T8 to L5. The posterior elements of hypoplastic L1 vertebra including the spinous process, bilateral lamina, transverse process, and both facet joints were removed. Then to performing VCD of deformed vertebra a pedicle probe was used to enlarge the pedicle holes on both sides to get enough access to the vertebral body then Cancellous bone of the vertebra body was removed through the pedicle holes by using a pituitary rongeur and a curette. To prevent the spinal cord injury due to unintentional vertebra displacement during VCD a temporary rod was inserted unilaterally. Then the posterior cortex of the deformed vertebral body was removed using a Kerrison rongeur and then an appropriate amount of the cancellous bone was removed to thin the anterior half of the vertebra which served as a hinge to correct the kyphotic deformity (figure 3). Then both rods were placed and by application of compressive force and osteoclasis of the anterior cortex the osteotomy site was closed. Then correction of deformity was confirmed by C-Arm fluoroscopy, the final fixation of instrumentation was completed. Then posterolateral fusion by combination of iliac bone graft and

allograft was performed then a drain was placed in the surgical site, and the wound was closed in layers.

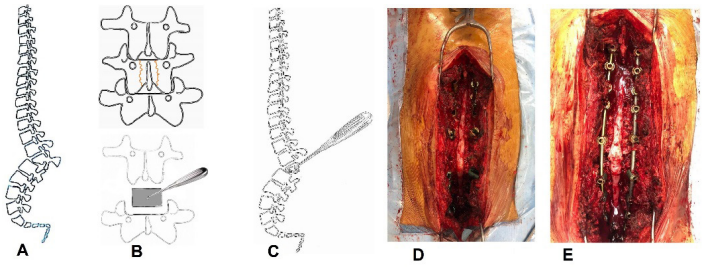

Figure 3: A.severe hypoplastic L1 vertebra . B-E: Laminectomy and Decancellation of vertebral body.

The whole blood loss was 500 ml. The surgical time was 6 hours. No early postoperative –h neurological complications or surgical site infection or CSF leak were reported. Patient was observed in the ICU overnight. He was allowed to sit in bed and stand with assistance on the first postoperative day. No postoperative bracing was used. The prophylactic antibiotic and drain were discontinued on postoperative day 2. The patient was discharged on postoperative day 5. The postoperative radiography showed acceptable coronal and sagittal alignment and head centered over the pelvis. T12-L2 segmental kyphosis angle was improved from 103 degrees to 42 degrees (Table1), and thoracic kyphosis (T5-T12) was improved from 17.5 degree to 52.3 degree, and lumbar hyperlordosis decreased from 81.5 degree to 54.5 degree. The postoperative SVA was +34mm.

Table 1: Summary of the radiographic results.

Corrective Ratios	Post-Operative	Pre-Operative	
59.2%	42	103	Segmental Kyphosis (T12-L2)
67.2%	56.8	18.6	Thoracic Kyphosis (T1-T12)
66.5%	52.3	17.5	Thoracic Kyphosis (T5-T12)
33.1%	54.5	81.5	Lumbar Lordosis (L1-S1)
43%	+34 mm	-45 mm	SVA

The clinical thoracolumbar kyphotic appearance improved after surgery (figure 4).

The neurological symptoms related to spinal stenosis improved substantially few weeks after surgery. His Gait disturbance improved remarkably. The patient was able to easily sleep in supine position. He returned to his job on postoperative day 30. VAS score was 8 preoperatively and 2 postoperatively which was improved about 43% (Table 2). ODI Score was 76% preoperatively and 13% postoperatively with 82.8% recovery rate. SRS-22 Score was 1.3 preoperatively and 4.8 postoperatively which was improved about 72.9%.

Table 2: Summary of VAS Score, ODI Score, SRS22 Score.

Recovery Rates	Post-Operative	Pre-Operative	
75%	2	8	VAS Score
82.8%	13%	76%	ODI Score
72.9%	4.8	1.3	SRS-22 Score

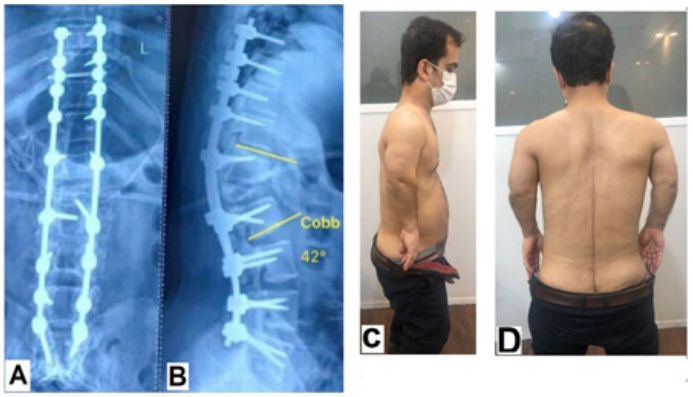

Figure 4: Post-operative radiographies and photos of the patient.

The patient was evaluated by radiography and physical examination every six months for 2 years. The follow-up radiography and CT scan showed acceptable spinal alignment and solid fusion and the patient had no back pain due to pseudoarthrosis or rod breakage during the 2 years of follow-up (figure 5).

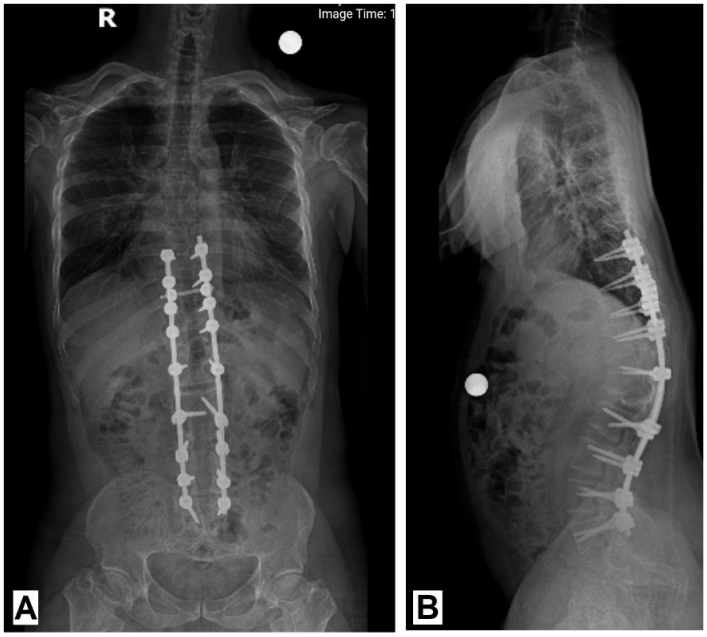

Figure 5: Follow up radiographies after 2 years.

Discussion

Thoracolumbar kyphosis usually is present at birth and progresses when the patients start to sit up and resolves spontaneously with improvement of muscle tones after standing and walking [1-5,11,17-28]. If the kyphosis progresses and exceeds greater than 30 degrees TLSO brace is recommended [1-5,8,10,29]. Surgery is required for progressive kyphotic deformity (more than 50 degree) and for those who develop significant neurological deficits [2,3]. Pedicle subtraction osteotomy or vertebral column resection [5,8,22,30,31] have been used for correction of achondroplastic thoracolumbar kyphosis but there are few reports about using Vertebral Column Decancellation (VCD) technique for these patients. Various osteotomies have been described for correction of spinal deformities. Schwab et al. classified spinal

osteotomy procedures into 6 grades [5]. For correction of small and flexible curves, Smith-Petersen osteotomies (Grade 1) or Ponte osteotomies (grade 2) may be sufficient. For more severe and rigid deformities, a pedicle subtraction osteotomy (Grade 3) or vertebral column resection (VCR) (grade 5) may be indicated. High grade osteotomies are technically challenging and a can be associated with a high rate of surgical complications. In PSO (Grade 3) the spinous processes and both lamina, pedicles, and a wedge shape bone of the posterior vertebral body are removed but adjacent disks remain intact. PSO has the limitation of correcting the rotational deformities. In VCR (Grade 5) the whole vertebral body and adjacent disks are resected and coronal, sagittal, translational, and rotational deformities can be corrected at the same time [5]. In Vertebral Column Decancellation (VCD) technique all the posterior vertebral elements including spinous processes and both lamina and pedicles are resected and then by a curette, the cancellous bone of vertebral body is removed and the cortical bones and adjacent disks remain intact [32,33]. VCD technique can correct larger angles than PSO, and compared with VCR, Vertebral Column Decancellation is technically simpler and safer with lower rate of complications [32,33]. Masato Tanaka et al. reported a case series of 3 patients with achondroplasia whit severe thoracolumbar kyphosis and neurologic deficits that underwent VCR. According to their results 66% of their patients had postoperative rod breakage that occurred at 1 and 2 years after surgery and 2 patients developed surgical site infection. They concluded that the results of VCR were acceptable for treating thoracolumbar kyphosis [8]. Wang et al. also reported a case series with 43% rod breakage during 18 months after VCR. Instability created by VCR procedure and pseudoarthrosis and impaired bone formation in achondroplastic patient in VCR procedure can be the cause of rod breakage [33,34]. Wenhao Hu et al. reported that Vertebral column decancellation is an effective treatment for correction of severe Pott's kyphosis [33]. Fanqi Hu and colleagues evaluated 31 patients with congenital kyphoscoliosis who underwent (AVCD) [32]. Their study demonstrated that AVCD can corrects spinal deformities in both the coronal and sagittal plane with satisfactory results and without additional neurological complications. In our patient who underwent Vertebral Column Decancellation (VCD) the preoperative segmental kyphotic angle (T12-L2) was 103 degrees and 42 degrees postoperatively with 59.2% correction rate. Thoracic kyphosis (T1-T12) was 18.6 degrees preoperatively and 56.8 degrees postoperatively with correction rate of 67.2% and thoracic kyphosis (T5-T12) was 17.5 degree preoperatively and 52.3 degree postoperatively with correction rate of 66.5%. The lumbar lordotic angle (L1-S1) was 81.5 degree before operation and 54.5 degree after operation with 33.1% correction rate. SVA was -45mm preoperatively and +34mm postoperatively with 75% correction rate. Vas score was 8 preoperatively and 2 postoperatively which was improved about 43%. ODI Score was 76% preoperatively and 13% postoperatively with 82.8% recovery rate. SRS-22 Score was 1.3 preoperatively and 4.8 postoperatively which was improved about 72.9%. Preoperatively the patient had severe back pain and neurological symptoms including numbness and claudication and after Vertebral Column Decancellation (VCD), back pain and numbness and claudication improved significantly. According to long term of the radiological and clinical evaluation we conclude that Vertebral Column Decancellation (VCD) procedure is an acceptable technique for management of thoracolumbar kyphosis in patient with achondroplasia.

Conclusion

The long term radiological and clinical results of Vertebral Column Decancellation (VCD) in treatment of sharp angular thoracolumbar kyphosis in achondroplastic mature patient demonstrated that this technique is a relatively safe procedure and also has favorable outcomes. Since surgical complications are the major concerns about 3-column osteotomies, strict adherence to agreed surgical protocols can minimize the rates of complications (such as dural rupture, surgical site infection, massive bleeding). With meticulous hemostasis and administration of tranexamic acid during the surgery the rate of blood loss can be reduced. Despite the impaired enchondral ossification in achondroplastic patients, due to the bone-to-bone contact in VCD, the risks of nonunion and subsequent complication such as rod breakage can be reduced.

Ethics approval

This study was conducted with approval from the Ethics Committee of Tehran University of Medical Sciences (TUMS) and was performed in accordance with the declaration of Helsinki.

References

1. Bridwell KH, Dewald RL. The textbook of spinal surgery. Lippincott-Raven Philadelphia PA. 1997.
2. Canale ST, Azar FA, Beaty JH, et al. Campbell's operative orthopaedics. Thirteenth edition. Elsevier Inc. Philadelphia PA. 2017.
3. Herring JA, Tachdjian MO Texas Scottish Rite Hospital for CHILDREN. Tachdjian's pediatric orthopaedics. Saunders/ Elsevier Philadelphia. 2008.
4. Rothman RH, Simeone FA, Herkowitz HN. Rothman-Simeone and Herkowitz's, the spine. Seventh edition 2018. Philadelphia Elsevier Philadelphia PA. 2018.
5. Winn HR. Youmans and Winn neurological surgery. Elsevier Philadelphia PA. Philadelphia PA. 2017.
6. Deng C, Wynshaw-Boris A, Zhou F, et al. Fibroblast growth factor receptor 3 is a negative regulator of bone growth. Cell. 1996; 84: 911-921.
7. Hasegawa K, Fukuhara R, Moriwake T, et al. A novel mutation p.Ser348Cys in FGFR3 causes achondroplasia. Am J Med Genet A. 2016; 170a: 1370-1372.
8. Tanaka M, Tsang Tung C, Haruo M, et al. Long-Term Results of Posterior Vertebral Column Resection for Severe Thoracolumbar Kyphosis with Achondroplastic Patients: A Case Series. Medicina (Kaunas). 2022; 58: 605.
9. Pauli RM. Achondroplasia: a comprehensive clinical review. Orphanet J Rare Dis. 2019; 14: 1.

10. Xu L, Li Y, Sheng F, et al. The Efficacy of Brace Treatment for Thoracolumbar Kyphosis in Patients With Achondroplasia. Spine (Phila Pa 1976). 2018; 43: 1133-1138.
11. Margalit A, McKean G, Lawing C, et al. Walking Out of the Curve: Thoracolumbar Kyphosis in Achondroplasia. J Pediatr Orthop. 2018; 38: 491-497.
12. Miyazaki M, Kanezaki S, Notani N, et al. Spondylectomy and lateral lumbar interbody fusion for thoracolumbar kyphosis in an adult with achondroplasia: A case report. Medicine (Baltimore). 2017; 96: 8983.
13. Qi X, Matsumoto M, Ishii K, et al. Posterior osteotomy and instrumentation for thoracolumbar kyphosis in patients with achondroplasia. Spine (Phila Pa 1976). 2006; 31: E606-610.
14. Eager M, Shimer A, Jahangiri FR, et al. Intraoperative neurophysiological monitoring (IONM): lessons learned from 32 case events in 2069 spine cases. Am J Electroneurodiagnostic Technol. 2011; 51: 247-263.
15. Neilipovitz DT, Murto K, Hall L, et al. A randomized trial of tranexamic acid to reduce blood transfusion for scoliosis surgery. Anesth Analg. 2001; 93: 82-87.
16. Pyeritz RE, Sack GH Jr, Udvarhelyi GB. Thoracolumbosacral laminectomy in achondroplasia: long-term results in 22 patients. Am J Med Genet. 1987; 28: 433-444.
17. Health supervision for children with achondroplasia. American Academy of Pediatrics Committee on Genetics. Pediatrics. 1995; 95: 443-451.
18. Ahmed M, El-Makhy M, Grevitt M. The natural history of thoracolumbar kyphosis in achondroplasia. Eur Spine J. 2019; 28: 2602-2607.
19. Bodensteiner JB. Neurological Manifestations of Achondroplasia. Curr Neurol Neurosci Rep. 2019; 19: 105.
20. Engberts AC, Jacobs WC, Castelijns SJ, et al. The prevalence of thoracolumbar kyphosis in achondroplasia: a systematic review. J Child Orthop. 2012; 6: 69-73.
21. Hashmi SS, Gamble C, Hoover-Fong J, et al. Multicenter study of mortality in achondroplasia. Am J Med Genet A. 2018; 176: 2359-2364.
22. Hunter AG, Bankier A, Rogers JG, et al. Medical complications of achondroplasia: a multicentre patient review. J Med Genet. 1998; 35: 705-712.
23. Khan BI, Yost MT, Badkoobehi H, et al. Prevalence of Scoliosis and Thoracolumbar Kyphosis in Patients With Achondroplasia. Spine Deform. 2016; 4: 145-148.
24. Kopits SE. Thoracolumbar kyphosis and lumbosacral hyperlordosis in achondroplastic children. Basic Life Sci. 1988; 48: 241-255.
25. Lutter LD, Longstein JE, Winter RB, et al. Anatomy of the achondroplastic lumbar canal. Clin Orthop Relat Res. 1977; 139-142.
26. Misra SN, HW Morgan. Thoracolumbar spinal deformity in achondroplasia. Neurosurg Focus 2003; 14: e4.
27. Pauli RM, Breed A, Horton VK, et al. Prevention of fixed, angular kyphosis in achondroplasia. J Pediatr Orthop. 1997; 17: 726-733.
28. Tanaka H. Achondroplasia: recent advances in diagnosis and treatment. Acta Paediatr Jpn. 1997; 39: 514-520.
29. Ando K, Kobayashi K, Nakashima H, et al. Sagittal alignment at 3 years old determines future thoracolumbar kyphosis in achondroplasia: A prospective study with minimum 5-year follow-up from infancy. N Am Spine Soc J. 2021; 6.
30. Ain MC, JA Browne. Spinal arthrodesis with instrumentation for thoracolumbar kyphosis in pediatric achondroplasia. Spine (Phila Pa 1976). 2004; 29: 2075-2080.
31. Wang Y, LG Lenke. Vertebral column decancellation for the management of sharp angular spinal deformity. Eur Spine J. 2011; 20: 1703-1710.
32. Hu F, Yang X, Wang C, et al. Asymmetrical vertebral column decancellation for the management of rigid congenital kyphoscoliosis. BMC Musculoskelet Disord. 2020; 21: 555.
33. Hu W, Zhang X, Yu J, et al. Vertebral column decancellation in Pott's deformity: use of Surgimap Spine for preoperative surgical planning, retrospective review of 18 patients. BMC Musculoskelet Disord. 2018; 19: 13.
34. Wang H, Wang S, Wu N, et al. Posterior Vertebral Column Resection (pVCR) for Severe Thoracolumbar Kyphosis in Patients With Achondroplasia. Global Spine J. 2022; 12: 1804-1813.