



# COMBINED, NOVEL MANAGEMENT OF BILATERAL VARUS HIP DEFORMITY USING “EIGHT-PLATE” IN CHILDREN WITH SPONDYLOEPIPHYSEAL DYSPLASIA CONGENITA

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**SUMMARY** – Spondyloepiphysal dysplasia congenita (SEDC) is a rare autosomal dominant genetic disorder. Femoral head ossification delay and the proximal femur varus deformity i.e. coxa vara (CV) are the major features of SEDC. The accepted treatment is a valgus femoral osteotomy. The data on hip surgery in SEDC are scarce. In our database from 2006 to 2020, there were 6 SEDC patients. Four patients had surgery on 8 hips. Surgical treatment was indicated due to progressive CV deformity i.e. a decreasing neck-shaft angle (NSA), pain, limited hip abduction, and gait disturbances. In three patients, a novel surgical treatment was applied – a greater trochanter apo-physiodesis using “Eight-plate”. The patients were evaluated clinically and radiologically. The median age at first surgery was 6.3 years (range, 3.2 to 9.5 y) and the median follow-up period was 7 years (range, 5.6 to 14 y). The postoperative NSA was significantly improved with a mean increase of 13 degrees ( $P < 0.001$ ). Additional surgeries were needed in two patients. Overall, our results showed improved clinical and radiological parameters. The purpose of this study was to determine whether a method using an “Eight-plate” applied early to greater trochanter apophysis in SEDC patients with bilateral CV could reduce the need for more aggressive surgery in near future.

**Keywords:** *Spondyloepiphysal dysplasia, hip, greater trochanter, surgery*

## INTRODUCTION

Spondyloepiphysal dysplasia congenita (SEDC) is a rare autosomal dominant genetic disorder characterised by type II collagen dysfunction. It has an incidence of 3-4 per million with clinical presentation varying from lethal to mild bone dysplasia<sup>1</sup>. The major features of SEDC are a femoral head ossification delay

and a proximal femur varus deformity i.e. coxa vara (CV)<sup>2-4</sup>. However, two types of CV were described in patients with SEDC. A milder type with no progressive CV, and another group of SEDC patients with a progressive CV with major disturbances in femoral head ossification and very short stature<sup>5</sup>.

Although the data on hip surgery in SEDC are scarce, the accepted treatment is a valgus femoral osteotomy<sup>4,6</sup>. This procedure is indicated due to a progressive natural history of the proximal femur varus deformity, a decreasing neck-shaft angle (NSA), pain, limited hip abduction, gait disturbances, and subsequent development of early hip osteoarthritis<sup>2-4</sup>.

The present study aims to report retrospectively analysed data obtained after surgical treatment of

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eight hips in four SEDc patients with the emphasis on the three patients treated initially as greater trochanter apo-physiodesis using “Eight-plate”. Therefore, we aimed to determine whether a minimally invasive method using an “Eight-plate”, applied early to greater trochanter apophysis in SEDc patients with bilateral CV, could reduce the need for more aggressive surgery in the future<sup>7</sup>.

## METHODS

All subjects were managed and observed at our institutions from 2006 to 2020 with a mean follow-up time of 7 years. The analysed data were collected from the medical records and other diagnostic tools used and medical history was taken from patients and their parents, Table 1. Four patients (3 females) at the age of 3 to 9 years, had surgery on 8 hips. The indication for surgical treatment was a progressive CV deformity i.e. a decreasing NSA, pain, limited hip abduction, and gait disturbances. In one patient at the age of 9.5 years, the surgical treatment was a proximal femur valgus osteotomy while in three female patients at the age of 3.2, 5.9, and 6.6 years respectively, a novel approach, i.e. guided growth principle with a greater trochanter apo-physiodesis using “Eight-plate”, was applied<sup>7</sup>.

The patients' medical history and preoperative clinical and radiologic data were reviewed and recorded. Radiological measurement included NSA as a measure of mechanical aspect and Hilgenreiner-trochanteric angle (HTA) as a measure to assess the CV deformity accurately in the presence of limited epiphyseal ossification<sup>8</sup>. Measurements were made before surgery and at the final follow-up. The difference was calculated. The parents of all four patients provided informed consent for publication.

## Surgical technique

The novel operative procedure, greater trochanter apo-physiodesis using “Eight-plate”, was performed by the senior author (D.A.) in three patients, while the valgus hip osteotomy was done in two patients by the senior author (D.A.) and the first author (J.V.). All patients were under general anaesthesia and placed supine with a linen insert under the ipsilateral buttock. In all surgeries, fluoroscopy was used to ensure proper plate and screw position. Using a direct lateral approach to the greater trochanter, after the fascia lata was longitudinally incised, “Eight-plates” were placed over previously inserted two K wires, one above, and the other below the greater trochanter growth plate. A lateral cortex was slightly drilled and screws were tightened creating greater trochanter apo-physiodesis Figure 1. Two patients had valgus hip intertrochanteric osteotomy on both hips in a standard fashion<sup>6</sup>. One patient was too old at the time of the first surgery to undergo greater trochanter apo-physiodesis with an “Eight-plate”. In this patient's valgus hip, osteotomy was secured with a standard 130° blade plate. In another patient, greater trochanter apo-physiodesis with an “Eight-plate” did not lead to satisfactory results and valgus hip osteotomy was secured with a Locking Cannulated Blade (LCB) Plate (OrthoPediatrics). However, because of the delayed healing at the osteotomy site and LCB plate breakage, revision surgery was needed on one hip and for this occasion, LCP Paediatric Hip Plate (Synthes) was used, Figure 2. During surgery, when the varus alignment is extreme, it is sometimes necessary to shorten the distal fragment (never more than 1 cm) to accommodate a good alignment of femoral fragments after the correction of the varus alignment is completed. Partial adductor tenotomy is sometimes added to femoral shortening.



*Figure 1. A) Intraoperative anterior-posterior fluoroscopy image of greater trochanter apo-physiodesis. “Eight plate” and screws are placed on the lateral side of the greater trochanter, inhibiting growth on the lateral side. B) Anterior-posterior X-ray image of properly placed “Eight plate” and screws at the lateral side of the greater trochanter on both hips. C) Anterior-posterior X-ray image of both hips in a patient treated with greater trochanter apo-physiodesis using “Eight plate”. While growing, the patient needed longer screws to obtain proper growth modulation.*

Table 1. Details of four patients included in this report.

Patient	Sex (M/F)	Age at first surgery (years)	First surgery	Second surgery	Third surgery	Fourth surgery	Follow up (years)
MS	M	9.5	Valgus hip intertrochanteric osteotomy	No	No	No	14
HE	F	6.6	Greater trochanter apo-physiodesis using "Eight-plate"	Longer screws in "Eight-plate"	No	No	7.4
AM	F	3.2	Greater trochanter apo-physiodesis using "Eight-plate"	No	No	No	6.6
LV	F	5.9	Greater trochanter apo-physiodesis using "Eight-plate"	"Eight-plate" removal from both hips	Valgus hip intertrochanteric osteotomy	Plate change in valgus hip intertrochanteric osteotomy	5.6

### Rehabilitation

All patients that underwent greater trochanter apo-physiodesis started with active and passive range-of-motion exercises from the first postoperative day. They were taught to use crutches for support and instructed to bear weight as tolerated. After three weeks, they were all able to walk without any assistance. However, patients that underwent valgus hip osteotomy had a slower recovery, and they used crutches for support for ten weeks.

### Clinical and radiological outcome measures

At the final follow-up visit, functional and clinical evaluation of the hips was done by assessing the patient's pain, range of motion, and gait. Radiological measurements included NSA and HTA.

### Statistical Analysis

Statistical analysis was performed by the statistical package Statistica. Statistics of descriptive data were calculated by standard formulas (median, range, minimum, and maximum). A paired-samples t-test was

performed to compare preoperative and postoperative NSA and HTA. A p-value <0.05 was regarded as statistically significant.

### RESULTS

A total of four patients, three girls, and one boy, with a median age of 6.3 years (range, 3.2 to 9.5 y) at the time of the first operation were included in the study. The patients' demographic data are listed in Table 1. The median follow-up was 7 years (range, 5.6 to 14 y). No intraoperative complications were reported. Two patients needed additional surgeries. Overall, the median NSA increased significantly from 117° (range, 102 to 127) preoperatively to 126° (range, 113 to 154) postoperatively (P<0.001). The median HTA increased significantly from -9° (range, -50 to 18) preoperatively to -2° (range, -23 to 22) postoperatively (P<0.001). However, for two patients treated only as greater trochanter apo-physiodesis using "Eight-plate" the median NSA increased significantly from 118° (range, 116 to 127)

Table 2. Preoperative and postoperative NSA and HTA change in surgically treated patients with spondyloepiphyseal dysplasia congenita for bilateral coxa vara. NSA = neck-shaft angle; HTA = Hilgenreiner-trochanteric angle.

Operated patients: greater trochanter apo-physiodesis using “Eight-plate”/ valgus hip osteotomy	NSA (°)				HTA (°)			
	preOP		postOP		preOP		postOP	
	RIGHT	LEFT	RIGHT	LEFT	RIGHT	LEFT	RIGHT	LEFT
MS	137	109	127	113	- 40	- 50	3	-22
HE	116	119	124	131	8	7	3	-6
AM	127	118	114	124	4	18	-23	-23
LV	110	102	150	154	-5	-11	19	22
Mean	117		130		-9		-3	

preoperatively to 124° (range, 114 to 151) postoperatively ( $P < 0.001$ ), while the median HTA values decreased significantly from 7° (range, 4 to 18) to -14° (range, -23 to 3) ( $P < 0.001$ ). The results of NSA and HTA patients' postoperative angle change are presented in Table 2. All patients reported clinical improvements such as pain decrease, greater range of motion, and better ability to walk.

## DISCUSSION

The main finding of this case series is that greater trochanter apo-physiodesis using “Eight-plate” for the treatment of bilateral CV in SEDc patients could improve the patient's clinical findings and NSA if applied early enough. Therefore, greater trochanter apo-physiodesis using “Eight-plate” may be valuable and beneficial for hips in SEDc patients, delaying or

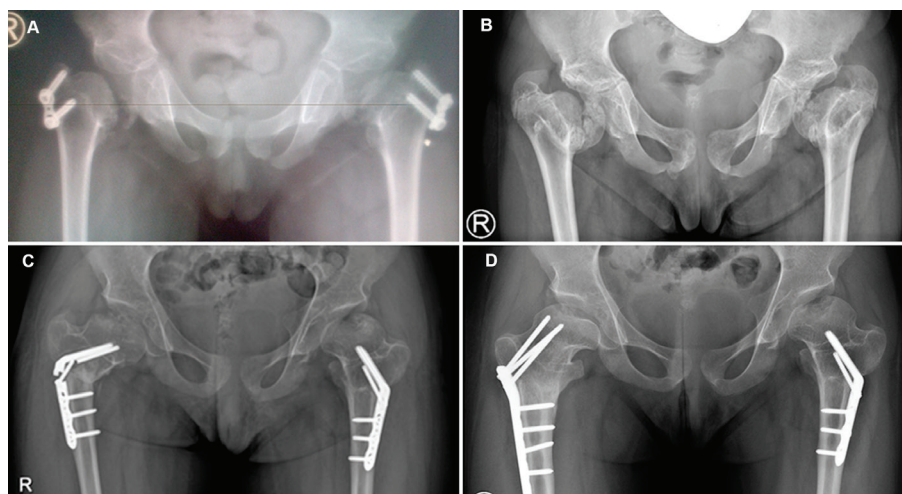


Figure 2. Anterior-posterior X-ray images of both hips in the same patient surgically treated for coxa vara. A) A greater trochanter apo-physiodesis using “Eight plate”. B) Unfavourable results after greater trochanter apo-physiodesis using “Eight plate”. There is still significant varus deformity, ie. intertrochanteric valgus osteotomy was indicated on both hips. C) Incomplete healing of the osteotomy on the right hip leads to implant breakage. Revision surgery was indicated. D) Last follow-up, both hips are with corrected neck-shaft angle. Intertrochanteric osteotomy on the right hip healed completely.

even obviating current alternatives such as valgus intertrochanteric osteotomy.

Because of a delayed and disrupted ossification of the capital femoral epiphysis in a SEDc patient, proper joint development is interrupted. The greater trochanter becomes relatively “overgrown” and prominent, reducing the effective abductor lever arm, limiting hip abduction, and causing gait disturbances. Moreover, the progressive natural history of the CV, pain and early hip osteoarthritis are major reasons why surgical treatment is indicated<sup>2-4</sup>.

In the present literature, there is a lack of studies describing the surgical treatment of CV in SEDc, with only two reports describing the proximal femoral valgus extension osteotomy using external fixation, or an internal plate fixation as a useful treatment option with improvements in hip pain, waddling gait, and hip range of motion<sup>2,4</sup>. Recently, it has been clearly emphasised that hip surgery for CV in SEDc patients is very challenging due to the three-dimensional deformity, unossified femoral head, and small patient size. Therefore, the use of complementary imaging modalities - computed tomography and magnetic resonance imaging was encouraged for preoperative planning<sup>9</sup>. Nevertheless, to the best of our knowledge, guided growth of the trochanteric apophysis for the treatment of CV in SEDc patients has never been reported.

Ever since Stevens PM in 2007 reported a technique of hemiepiphysiodesis with an “Eight-plate” and screws using a tension band principle over the growth plate, due to favourable results, its application has become very popular for various alignment corrections such as tibia vara or valgus of the ankle joint<sup>7,10,11</sup>. In addition, the same author described its application for guided growth of the trochanteric apophysis suggesting that tethering the greater trochanteric physis in combination with a medial soft tissue release, could avoid osteotomy management in the treatment of Legg-Calve-Perthes disease<sup>12</sup>. Moreover, Stevens used the guided growth principle of the trochanteric apophysis in three patients with chondrodysplasia for treatment of CV hoping to postpone or avert intertrochanteric osteotomy<sup>13</sup>. However, reasonable arguments were raised by other authors questioning the balancing between the rebound phenomenon and continuing natural angle changes at the hip after guided growth were applied for the treatment of displaced hips<sup>14</sup>.

In our study, a greater trochanter apo-physiodesis with an “Eight plate” was applied in three patients that

were all below the age of seven at the initial surgery, Table 1. This surgical method should not be used before the two years of age since children’s walking activities are not frequent and strong enough to produce a progression of CV deformity. After such treatment, in two of our patients there was no need for valgus intertrochanteric osteotomy at the most recent follow-up, while one patient underwent valgus intertrochanteric osteotomy on both hips. Due to a plate breakage, revision surgery was needed on one hip, Figure 2. All our patients had clinical improvements after surgical treatment in terms of pain relief, range of motion increase, and waddling gait correction. However, in two patients treated only with greater trochanter apo-physiodesis radiological measurement of NSA improved on three hips, while HTA decreased on all four hips, Table 2. Based on our findings and the recent report by Bayhan et al, it is not clear to determine whether a HTA in patients with skeletal dysplasia, as described by Oh et al. is a suitable radiological parameter to use when clinically assessing SEDc patients postoperatively<sup>4,8</sup>. On the other hand, NSA improved on three hips out of four, and this parameter seems to be more reproducible. However, in patients with SEDc ossification of the femoral head and neck is often delayed or even absent. Therefore, for these occasions, it is difficult to use traditional measurements to assess the severity of the proximal femoral deformity<sup>8</sup>.

Proximal femoral valgus osteotomy has been described for different aetiologies of CV. Shetty et al. and more recently Bayhan et al. reported surgical correction of CV in SEDc patients by proximal femoral valgus osteotomy as an effective treatment that improved hip pain and range of motion whether an external fixator or internal plate was used<sup>2,4</sup>. While Shetty et al. used a hybrid external fixator to perform proximal femoral valgus-extension osteotomy with distal femoral varus osteotomy in eight SEDc patients, Bayhan et al. performed valgus extension osteotomy of the proximal femur on 48 hips in 26 children with the average age of 9.6 at the time of surgery and the average age of 5 at the follow-up<sup>2,4</sup>. In our two patients treated with valgus intertrochanteric osteotomy on both hips, significant clinical improvements were noted. Besides clinical, significant radiological measurement improvements of both observed angles were noted, Table 2. This might imply that NSA and HTA are more reproducible radiological measurements in SEDc patients who had undergone valgus intertrochanteric osteotomy than in

those patients where greater trochanter apo-physiodesis with "Eight-plate" was done.

According to our results, greater trochanter apo-physiodesis with "Eight-plate" corrected CV in two SEDc patients on three hips. The ossification of the capital femoral epiphysis that was observed during follow-up might suggest a positive effect of the procedure on hip development. However, it is very challenging to conclude whether femoral head ossification is a result of the natural history or the greater trochanter apo-physiodesis with "Eight-plate". Nevertheless, because of low morbidity, feasible surgical technique, and low costs, we believe that greater trochanter apo-physiodesis with "Eight-plate" is a new valuable method for the first-line treatment of CV in SEDc patients below the age of seven. The mean follow-up of our patients is 7 years, and at that time there was no need for salvage procedures, like pelvic osteotomies which are suggested for major deterioration<sup>4</sup>. However, since at initial surgery, the average age was 6.3 years, a longer follow-up and further comparative studies are needed to validate the broad applicability of this novel approach and to document its long-term outcome in comparison to currently used strategies.

The limitation of this study is the low number of patients; therefore, we cannot give definitive remarks regarding the efficacy of the greater trochanter apo-physiodesis using "Eight-plate" for the treatment of CV in SEDc patients. However, we believe that the present study is justified based on the rarity of SEDc, as well as on the mid-term follow-up. The strength of this study is that all greater trochanter apo-physiodesis using "Eight-plate" procedures were done by a single orthopaedist. Therefore, an accumulated and fine-tuned experience with this novel surgical approach in such a rare condition as SEDc is presented.

In light of previous statements, it can be concluded that the mid-term results of the greater trochanter apo-physiodesis using "Eight-plate" is a minimally invasive and viable surgical treatment option for the treatment of bilateral CV in SEDc patients. If this technique is applied early enough - before the age of seven, there is a greater chance that it will achieve its major advantage - a decrease of the need for more extensive surgery in the future. However, a close patient follow-up and a full knowledge of valgus intertrochanteric osteotomies is needed to provide the best possible care for SEDc patients. In addition, the fact that this study was done using a guided growth of the

greater trochanter method in combination with NSA and HTA analysis for bilateral CV assessment in three SEDc patients contributes to future research. This can serve as a template for future multicentre collaborative studies, as it provides an objective measure for more accurate, longer-term evaluation.

## DECLARATION OF COMPETING INTEREST

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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#### Sažetak

### NOVI NAČIN LIJEČENJA BILATERALNOG VARUS DEFORMITETA KUKA KORIŠTENJEM "PLOČICE OSMICE" KOD DJECE S KONGENITALNOM SPONDILOEPIFIZEALNOM DISPLAZIJOM

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Spondiloepifizna displazija kongenita (SEDC) je rijedak autosomno dominantan genetski poremećaj. Glavna značajka SEDC je odgoda okoštavanja glave bedrene kosti i varusna deformacija proksimalnog femura, tj. coxa vara (CV). Prihvaćeno liječenje je valgus osteotomija proksimalnog femura. Podaci o operaciji kuka u SEDC pacijenata su oskudni. U našoj bazi od 2006. do 2020. bilo je 6 SEDC pacijenata. Četiri pacijenta imala su 8 operacija kukova. Indikacija za kirurško liječenje bila je progresivna CV deformacija tj. smanjenje vratno-dijafizičnog kuta (NSA), bol, ograničena abdukcija kuka i poremećaji hoda. Kod tri pacijenta primijenjeno je novo kirurško liječenje - apofiziodeza velikog trohantera pomoću "pločice osmice". Bolesnici su praćeni klinički i radiološki. Medijan dobi kod prve operacije bio je 6,3 godine (raspon od 3,2 do 9,5 godina), a medijan praćenja 7 godina (raspon od 5,6 do 14 godina). Postoperativni NSA je značajno poboljšana s prosječnim povećanjem od 13 stupnjeva ( $P < 0,001$ ). Dva pacijenta su trebala dodatne operacije. Sve u svemu, naši su rezultati pokazali poboljšane kliničke i radiološke parametre. Svrha ove studije bila je utvrditi može li metoda koja koristi "pločicu osmicu", rano primijenjena na apofizu velikog trohantera kod SEDC pacijenata s obostranom CV-om, smanjiti potrebu za ekstenzivnijom operacijom u bliskoj budućnosti.

**Ključne riječi:** *Spondiloepifizna displazija, kuk, veliki trohanter, operacija*