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# Inverted V-shaped high tibial osteotomy for severe tibia vara associated with Turner syndrome: A case report and review of literature

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## ABSTRACT

**INTRODUCTION:** Severe cases of genu varum represent a major challenge in obtaining normal configuration of the proximal tibia and overall limb alignment.

**PRESENTATION OF CASE:** We performed inverted V-shaped high tibial osteotomy (HTO) by using a locking plate for recurrent severe bilateral tibia vara in a 15-year-old female patient with Turner syndrome. Preoperative medial proximal tibial angle (MPTA) and standing femorotibial angle (FTA) of the right/left legs were 67°/69° and 197°/203°, respectively. In order to obtain overall neutral alignment, the correction angle in the right/left knees was required to be 23°/32°. Preoperative planning demonstrated that inverted V-shaped HTO could provide sufficient correction angle with large bone stock and wide bony contact. A postoperative full-standing radiograph showed that the mechanical axes passed through the center of right/left knees with 87°/88° of MPTA.

**DISCUSSION:** Inverted V-shaped HTO has advantages, as it requires a smaller amount of bone resection and smaller opening gap compared to the closing-wedge and opening-wedge osteotomies.

**CONCLUSION:** Inverted V-shaped HTO can be a useful surgical method to treat severe tibia vara in order to obtain adequate configuration of the proximal tibia and overall limb alignment.

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## 1. Introduction

Turner syndrome is caused by the absence of total or part of the X chromosome and have various anatomic anomalies including in the musculoskeletal system. Turner syndrome can cause progressive varus deformity of the proximal tibia because of premature epiphyseal closure in the medial proximal tibia [1]. Severe cases of genu varum represent a major challenge in obtaining normal configuration of the proximal tibia and overall limb alignment [2]. Several types of osteotomies have been described for the correction of angular deformity of the knee. Among them, inverted V-shaped high tibial osteotomy (HTO), which was proposed in 1973 by Levy et al. [3], is advantageous, especially in cases of severe deformity, by providing sufficient correction angle with large bone stock and wide bony contact [4].

We applied inverted V-shaped HTO to correct severe bilateral tibia vara in a patient with Turner syndrome, which had recurred after corrective proximal tibial osteotomy below the growth plates. Few reports are available regarding the effectiveness of inverted

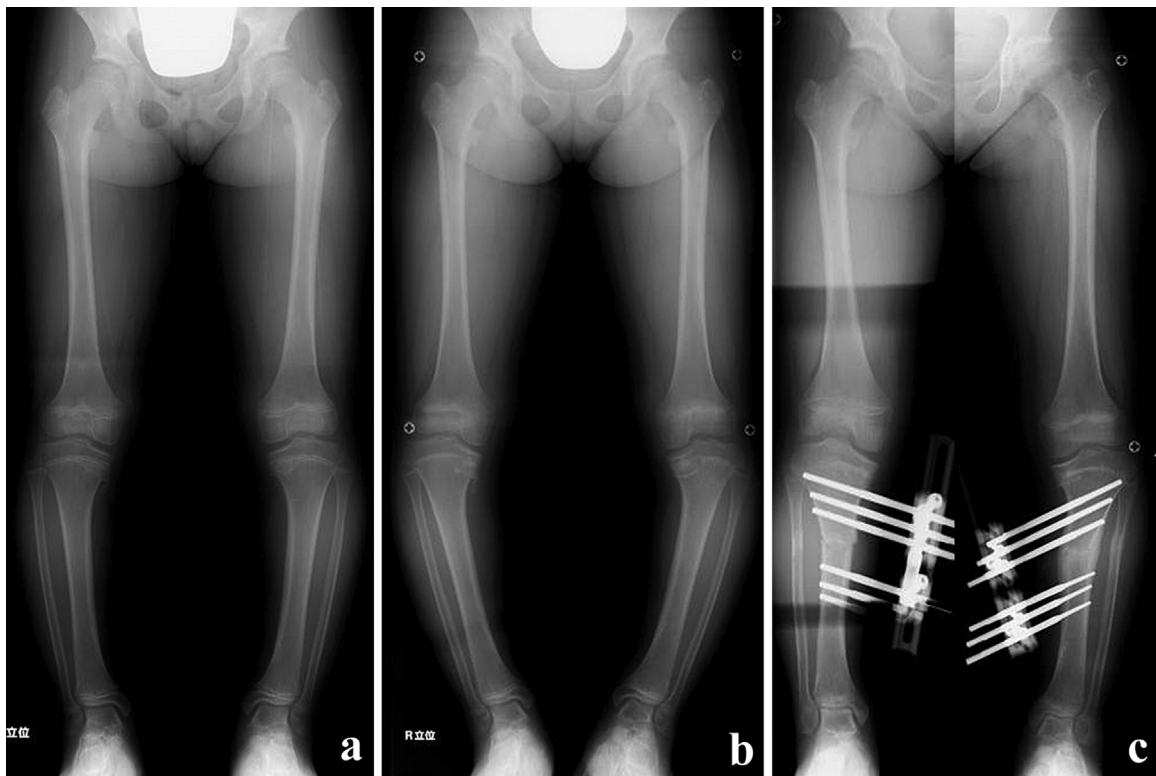
V-shaped HTO for severe proximal tibial varus deformity. Therefore, we report our clinical experience with the application of this surgical technique together with a literature review.

## 2. Case report

The patient was diagnosed with Turner syndrome based on chromosome test results at the age of 8 months and started receiving growth hormone therapy at the age of 4 years. She was referred to our department at the age of 10 years for bilateral genu varum (Fig. 1a) along with a short neck, barrel chest, cubitus valgus, Madelung deformity, and short fourth metacarpal. She had no limited range of limbs motion, joint laxity, limb length discrepancy, other complaints, or medical comorbidities. Full-standing radiographs showed progressive tibia vara over a two-year follow-up period due to premature epiphyseal closure of the medial proximal tibia with bony bridging across the physes (Fig. 1b). She underwent acute valgus osteotomy of the proximal tibial shaft by using an external fixator at the age of 12 years (Fig. 1c). Bilateral genu varum recurred subsequent to the osteotomy of the proximal tibial shaft (Fig. 2). At the age of 15 years, medial proximal tibial angle (MPTA) and standing femorotibial angle (FTA) in the right/left legs were 67°/69° and 197°/203°, respectively. Normal ranges for the

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**Fig. 1.** Long-leg standing preoperative radiographs showing bilateral tibia vara at the age of 10 years (a) and progressive deformity at the age of 12 years (b). Corrective osteotomies of the proximal shaft of both tibias were performed with an external fixator at the age of 12 years (c).

MPTA and FTA were reported to be  $85^{\circ}$ – $90^{\circ}$  and  $173^{\circ}$ – $178^{\circ}$ , respectively [5]. Mechanical lateral distal femoral angle (mLDFA) in the right/left legs were maintained at  $85^{\circ}/85^{\circ}$ , indicating normal alignment:  $85^{\circ}$ – $90^{\circ}$ , and lateral views of the knees did not show any significant deformity. Transepiphyseal osteotomy was planned to correct the severe tibial deformity and prevent future recurrence underlying lateral open physis.

Surgical options included, a closing-wedge HTO, opening-wedge HTO, and inverted V-shaped HTO (Fig. 3). Elevation of the medial plateau was not considered because of normal congruity of the joint surfaces and knee stability [6]. On preoperative planning, closing-wedge HTO showed marked morphological change and decreased bone stock of the proximal tibia (Fig. 3a). Opening-wedge HTO demonstrated too large of an opening gap on the medial side to obtain sufficient correction angle (Fig. 3b). Compared with these other procedures, inverted V-shaped osteotomy would require a smaller amount of bone resection and opening gap (Fig. 3c). Thus, in this case, we performed consecutive inverted V-shaped HTOs by using polyaxial locking plates (NCB PT<sup>®</sup>, Zimmer Inc., Warsaw, IN, USA), with correction angles of  $23^{\circ}/32^{\circ}$  in right/left knees, respectively.

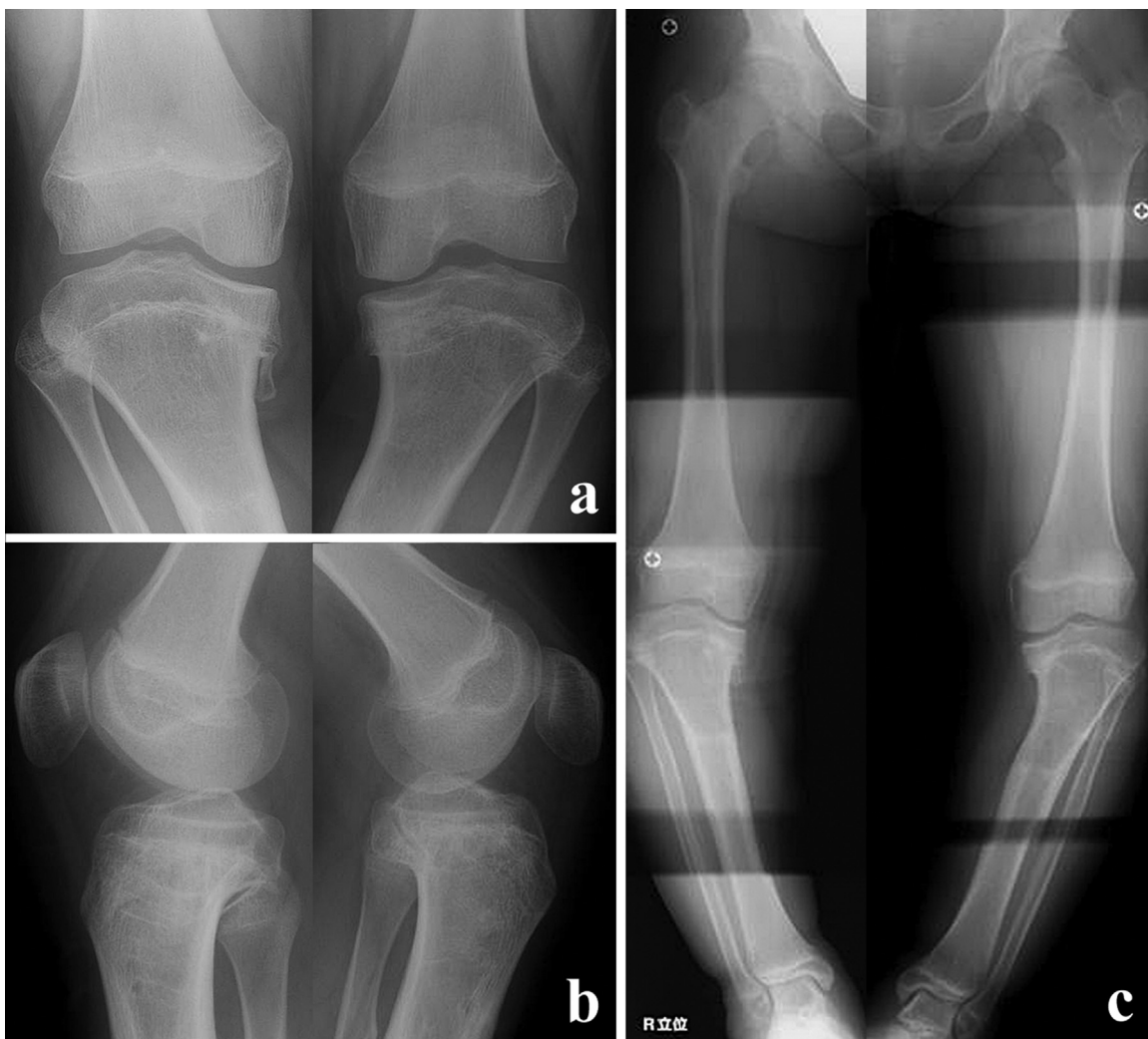
The procedure was performed by senior surgeons according to the previously described methods [3,4]. Briefly, the fibula was osteotomized in the middle third [7]. The anterior and posterior cortices and periosteum at the apex of the V shape were preserved and used as a central hinge during correction by applying valgus stress (Fig. 4). Distal locking screws were placed using minimally invasive plate osteosynthesis (MIPO) technique. Postoperative radiograph showed that mechanical axes pass through the centers of the knees, with  $174^{\circ}/172^{\circ}$  of standing FTA and  $86^{\circ}/88^{\circ}$  of MPTA in right/left legs (Fig. 5a–c). Knee exercises with a passive range of motion and partial weight-bearing were started five days and three weeks after surgery, respectively, as formal physiotherapy. She completed growth hormone therapy and underwent the

plate extraction at the age of 17.5 years. At more than 1.5 years after the inverted V-shaped osteotomies, she demonstrated bilaterally adequate configuration of the proximal tibia with residual right distal tibia varus and left mid-tibia valgus (Fig. 5d) and no functional limitations during daily activity. This manuscript has been reported in accordance with the SCARE criteria [8].

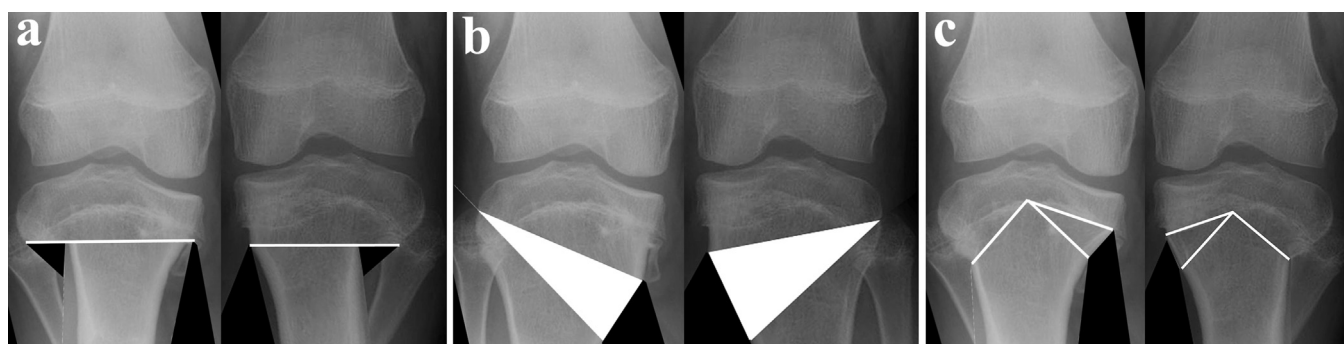
### 3. Discussion

Inverted V-shaped HTO was successfully performed for severe bilateral tibia vara in a 15-year-old female patient with Turner syndrome. Both right and left knees required a large correction angle exceeding  $20^{\circ}$  ( $23^{\circ}$  and  $32^{\circ}$ , respectively). However, restoration of normal limb alignment and joint-line obliquity ( $86^{\circ}/88^{\circ}$  of MPTA, respectively) [9] was achieved to prevent progression toward secondary knee osteoarthritis. Furthermore, acute osteotomy with the use of polyaxial locking plates contributed to the rapid recovery and early return to normal function.

Leg deformities in patients with Turner syndrome comprise genu valgum (knock-knee) in 60% of cases [10]. Although it is rare, Turner syndrome is one of the causes of progressive bilateral tibia vara [1]. Irreversible medial epiphyseodesis due to genetic predisposition and asymmetrical pressure on the physis has been recognized to induce a relatively high rate of recurrence of varus deformity after proximal tibial osteotomy [11–15]. Therefore, in this case, Langenskiöld's procedure consisting of bone bridge resection and free fat grafting [16], permanent hemiepiphysiodesis [17], or hemiepiphyseal eight-Plating [18] should be considered in addition to the acute valgus osteotomy of the proximal tibial shaft by using an external fixator at the age of 12 years. On this occasion, transepiphyseal HTO combined with locking plate fixation was chosen to close the lateral epiphysis permanently and prevent recurrent varus deformity. Then, different types of procedures, including closing-wedge, opening-wedge, and inverted V-shaped



**Fig. 2.** Radiographs at the age of 15 years showing recurrence of the severe tibia vara in the right/left limb with 66°/69° of the medial proximal tibial angle and 194°/192° of the standing femorotibial angle.

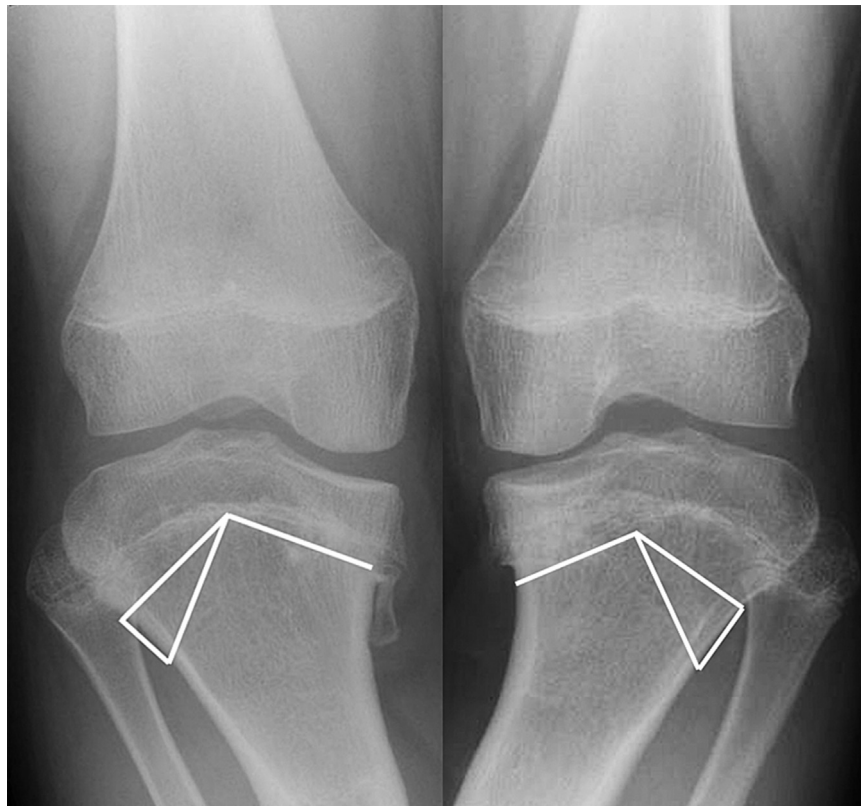


**Fig. 3.** The surgical procedures considered that the mechanical axis passed through the centers of the knees. In the closing-wedge high tibial osteotomy (HTO), the proximal tibial osteotomy level is set at 2 cm below the medial joint line and the lateral bone wedge was removed depending on the correction angle (a). In the opening-wedge HTO, the proximal tibial osteotomy level is set at 3.5 cm below the medial joint line, running obliquely upward to the tip of the fibular head (b). In the inverted V-shaped osteotomy, the apex of the “V” is proximal to the tibial tuberosity under the patellar ligament (c).

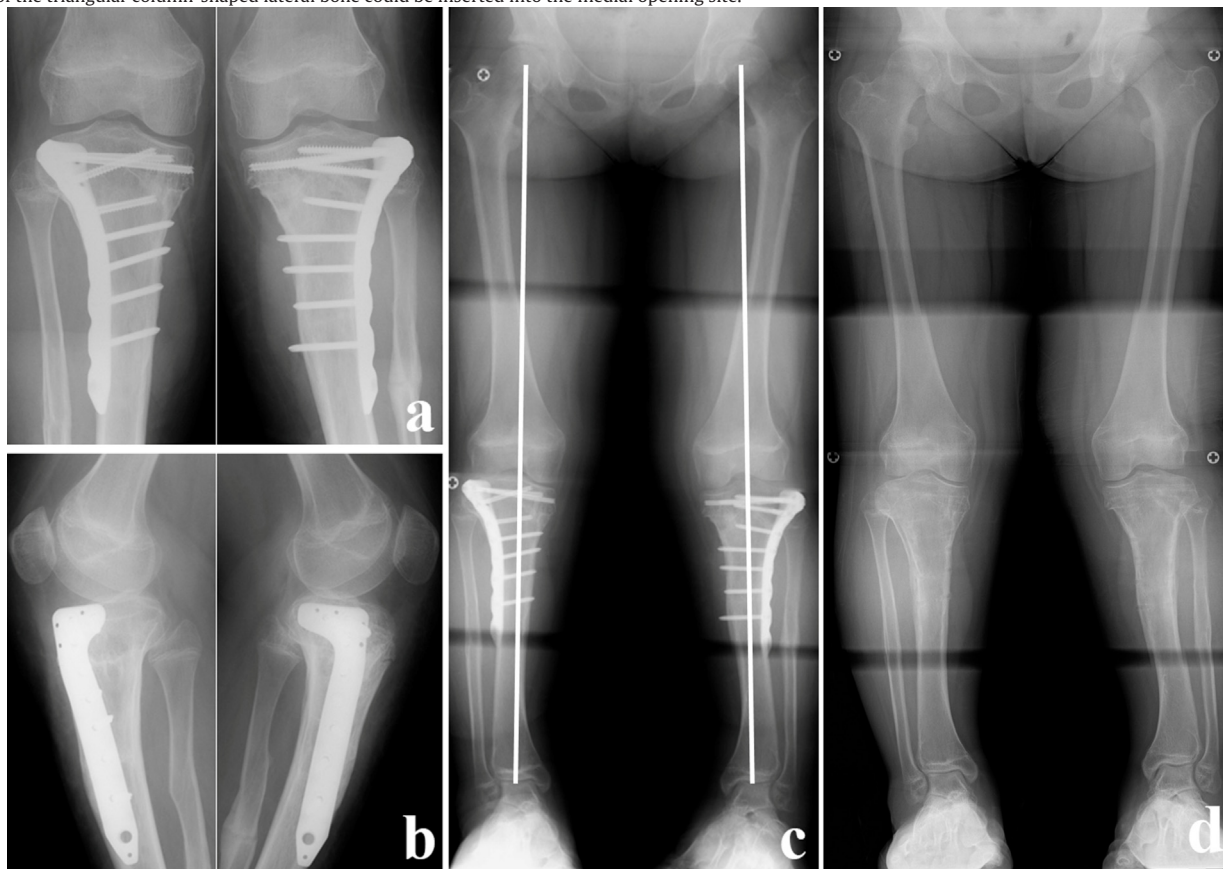
HTO, became variable surgical treatment options to obtain neutral overall alignment and joint-line obliquity.

Preoperative planning and operative technique selection are essential to obtain neutral overall alignment in severe tibia vara. Inverted V-shaped HTO has advantages, as it requires a smaller amount of bone resection and smaller opening gap compared to the closing-wedge and opening-wedge osteotomies. Wide bony con-

tact and a polyaxial locking plate provide not only a stable fixation at the osteotomy site but also early bone union, even in cases with a large correction angle [4]. Gradual correction with the use of an external fixator is another surgical treatment option with the ability to tune correction postoperatively [1,13]; however, its significant disadvantage is the need for prolonged external fixation [19]. We therefore applied inverted V-shaped HTO in this case and achieved



**Fig. 4.** Preoperative planning for the inverted V-shaped osteotomy demonstrates that right and left knees required correction angles of 23° and 32°, respectively. The resected segment of the triangular column-shaped lateral bone could be inserted into the medial opening site.



**Fig. 5.** Postoperative radiographs showing complete valgus correction; the mechanical axis passes through the center of the knees; the MPTA is 87°/88° (a, b), and the standing FTA is 174°/172° in the right/left legs, respectively (c). Full-standing radiographs at the most recent follow-up showing retained bilaterally adequate configuration of the proximal tibia with residual right distal tibia varus and left mid-tibia valgus (d).

successful acute correction of severe bilateral tibia vara with quick recovery to normal function. Disadvantages of this procedure are the need for fibular osteotomy, more extensile approach with soft tissue dissection and stripping than a typical closing-wedge and opening-wedge HTOs, and precise intraoperative evaluation of the corrected limb alignment.

In inverted V-shaped HTO, the center of rotation is the apex of the V shape, which is proximal to the tibial tuberosity under the patellar ligament and could have little impact on patellar height and mediolateral shift of the proximal tibia [3,4]. El-Azab et al. reported that closing-wedge HTO leads to an increase in patellar height, whereas the incidence of patella infera increases after opening-wedge HTO [20]. Altered patellar height could induce anterior knee pain and result in long-term patellofemoral cartilage degeneration [21]. Furthermore, residual proximal tibial deformity affects the degree of difficulty in possible future total knee arthroplasties [22]. Right distal tibia varus and left mid-tibia valgus at final follow-up in this case could be due to the skeletal abnormalities of Turner syndrome and previous valgus osteotomy of the tibial shaft.

#### 4. Conclusions

Inverted V-shaped HTO can be a useful surgical method to treat severe genu varum associated with Turner syndrome in order to obtain adequate configuration of the proximal tibia and overall limb alignment. Long-term follow-up is required to know that the final distal deformities may affect the patient's ankle and overall function in the future and may accelerate development of osteoarthritis.

#### Conflicts of interest

All authors have no potential conflicts of interest to disclose.

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#### Ethical approval

Ethical approval is not applicable to case report in our institution.

#### Consent

Written informed consent was obtained from the parents for the publication of this case report and accompanying images.

#### Author contribution

DA, SH, MY, and TK were the physician involved in the care of the patient. DA and SH were the authors of the article. KO, YN were the supervising physicians in the care of the patient. All authors read and approved the final manuscript.

#### Registration of research studies

N/A.

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#### Guarantor

Satoshi Hamai

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