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Case Report

# Staged Bilateral Total Hip Arthroplasty in a Patient With Larsen Syndrome

Joseph X. Robin, MD *, Nathan Huebschmann, BA, Jordan Villa, MD, Ran Schwarzkopf, MD

*Division of Adult Reconstruction, Department of Orthopedic Surgery, NYU Langone Health, New York, NY, 10003, USA*

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ABSTRACT

Larsen syndrome is a rare genetic disorder characterized by weak connective tissues and various musculoskeletal abnormalities. This is a case report of a 39-year-old patient with Larsen syndrome who presented with over a decade of bilateral hip pain and difficulty ambulating. This patient has a prior history of bilateral congenital hip dislocations that were treated with open reduction and spica casting as a child with good result. Years later, she went on to develop bilateral hip osteoarthritis with significant remodeling of the proximal femur. The goal of this case presentation is to demonstrate the utility of total hip arthroplasty for this patient and discuss surgical challenges and considerations.


## Introduction

Larsen syndrome is a rare genetic disorder with an estimated incidence of 1 in 100,000 live births [1]. The syndrome was first described by Larsen and colleagues in a series of 6 children in 1950 [2]. In this series, they observed a pattern of bilateral dislocations of the hips, knees, and elbows along with craniofacial abnormalities, skeletal abnormalities of the feet and fingers, and occasional palate or spinal segmentation abnormalities. The description of additional cases in the literature have corroborated the characteristic features of Larsen syndrome to be multiple dislocations of the large joints [3-5]; craniofacial anomalies [3,6]; cervical kyphosis and scoliosis [4,7-10]; and skeletal anomalies such as clubfoot and long, cylindrical, or spatulate fingers [3-5]. Given the potential for cervical spine dislocation, cardiovascular problems (eg, interventricular or inter-atrioventricular communication), and respiratory complications, Larsen syndrome can be fatal at an early age [4,11].

Two forms of Larsen syndrome have been described based on genetic etiology. The classic form of Larsen syndrome is caused by clustered missense mutations in the filamin B gene coding for the filamin B cytoskeletal protein and is typically inherited in an autosomal dominant manner [12]. The other form of Larsen syndrome, referred to as autosomal recessive Larsen syndrome, is caused by mutations in the carbohydrate sulfotransferase 3 gene and shares phenotypic features with humero-spinal dysostosis and spondyloepiphyseal dysplasia Omani type [13].

Although there are classical features of Larsen syndrome, its presentation can be heterogenous. Hip pathology has been reported in approximately 60% of cases in the literature. Hip-specific problems include subluxation and dislocation due to capsuloligamental hyperlaxity [4]. A recent case series described bilateral one-stage total hip arthroplasties (THAs) performed in 2 siblings with severe end-stage bilateral arthritis secondary to Larsen syndrome [14]. Two-year follow-up revealed improvement in function and quality of life, suggesting THA may be a viable option for patients with Larsen syndrome and resultant hip arthritis.

The present case report describes a patient with Larsen syndrome with a prior history of congenital bilateral hip dislocations treated surgically in childhood who underwent staged bilateral THA. THA following prior procedures for instability carries a significant risk of postoperative instability and/or other mechanical failures requiring possible revision surgery [15,16]. The purpose of this report is to describe the indications for THA in patients with Larsen syndrome, operative challenges, and postoperative course. The patient provided written informed consent that her case would be submitted for publication.

## Case history

A 39-year-old female (weight, 45 kg; height, 116 cm; body mass index, 33.2 kg/m2) with Larsen syndrome presented to the office

* Corresponding author. Division of Adult Reconstruction, Department of Orthopedic Surgery, NYU Langone Health, 301 East 17th Street, New York, NY 10003, USA. Tel.: +1 205 908 6889.
*E-mail address:* Joseph.Robin@nyulangone.org

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with several years of bilateral hip pain, left greater than right. The patient was born with bilateral hip dislocations that were treated successfully with greater trochanteric osteotomy, open hip reduction, and spica casting in childhood. Later in childhood, she developed progressive scoliosis that was treated with a thoracolumbar spine fusion extending from T3 to L5. The patient also had multiple bilateral knee procedures related to tibiofemoral joint incongruity with acquired bilateral knee subluxations.

Initially, the patient presents with a chief complaint of left hip pain. She ambulates with great difficulty using bilateral axillary crutches. She complains of left hip pain localizing to the groin and the lateral aspect of her hip that radiates to the anterior thigh and knee. She describes the pain as constant, and exacerbated with physical activity including standing from a chair, walking, putting on socks and shoes, and ascending and descending stairs. She also complains of pain at rest that wakes her from sleep. She reports that the pain and stiffness have become functionally debilitating, resulting in an overall loss of functional independence.

Clinically, she has a well-healed surgical scar over the lateral aspect of bilateral hips and no perceived leg length discrepancy. On physical exam, she has no tenderness about the hips and pain with log-roll and heel-strike. Her left hip range of motion is 0 to 70 degrees of flexion, 0 degrees of internal rotation, 15 degrees of external rotation, and 15 degrees of abduction and adduction. Her right hip range of motion is 20 to 70 degrees of flexion, 0 degrees of internal rotation, 15 degrees of external rotation, 15 degrees of abduction, and 5 degrees of adduction. Her hips are stable through the range of motion, but she has significant pain at the extremes of range of motion.

The preoperative radiographs (Fig. 1) reveal severe bilateral hip arthrosis with global bone-on-bone osteoarthritis. She has bilateral protrusio acetabuli with increased lateral coverage of the femoral head. She has bilateral well-healed trochanteric osteotomies, and her left hip has a short valgus neck and collapsed femoral head, while her right hip has more pronounced femoral and acetabular osteophytes but less femoral head collapse. Standing full-length radiographs (Fig. 2) demonstrate multilevel cervical spine degenerative disc disease and facet arthropathy, a dextroconvex lumbar curvature with compensatory left-sided pelvic obliquity. On the lateral standing radiographs, this patient is noted to have well-balanced sagittal alignment characterized by a high pelvic incidence and appropriate lumbar lordosis in her rigid fusion above.

In addition, on the lateral standing radiograph, this patient is noted to have an anterior pelvic tilt that changed by 20 degrees on the lateral sitting radiograph. The sacral slope is 47 degrees standing and 29 degrees sitting.

Given the patient's persistence of pain and loss of function despite nonoperative management including physical therapy, ambulatory assist devices, corticosteroid injection, and non-steroidal anti-inflammatory drugs, the patient was indicated for left THA.

We used the posterior approach with the aid of computer navigation (Intellijoint HIP, Waterloo, Canada). A 40 millimeter (mm) multihole acetabular shell (Trident PSL, Stryker, Kalamazoo, MI) and a 15 mm screw were placed. The acetabular component was placed in 43 degrees of inclination and 32 degrees of anteversion. Due to the small diameter of the patient's femoral diaphysis (12 mm proximally and 7 mm at mid-diaphysis) a burr (2.3 × 22 mm tapered router, Stryker, Kalamazoo, MI) was used to create the lateral entry point in order to effectively cannulate the femur. The femoral canal was then reamed by hand until rigid endosteal contact was obtained. An uncemented, 13 mm, 135-degree monoblock tapered and fluted stem was used (Wagner Cone, Zimmer-Biomet, Warsaw, IN). A prophylactic 2 mm cable was placed below the level of the lesser trochanter (Stryker, Kalamazoo, MI). The bearing surface was a neutral-faced, highly crossed polyethylene liner and a 28 mm plus 0 ceramic head (Zimmer-Biomet, Warsaw, IN) (Fig. 3). The hip was then ranged through various positions to ensure adequate stability.

Postoperatively, the patient recovered well and was discharged home on postoperative day 1. Her left hip pain drastically improved, and she was able to ambulate well with a rolling walker. The patient resumed working with physical therapy and started to notice that her right hip was causing great pain and limiting her progress. At 6-months postoperatively, the patient returned to the operating room for the right THA, which proceeded in a similar fashion with no complications (Fig. 4).

Following the right THA, the patient recovered well and was discharged home. Postoperatively, she has done well and has had an excellent result at 1 year follow-up. The patient continues to use a rolling walker when out of the house to aid in balance, but she has resumed an active lifestyle. She walks several blocks per day and reports no hip pain.

## Discussion

THA is a highly successful operation for generating pain relief and improving functional status, all while maintaining a low risk of complications. Patients such as the 1 above present a challenge for many reasons.

First, this patient was instructed by other providers in the past that she was too young for a THA and therefore waited for several years, resulting in years of pain and functional decline. While THA is

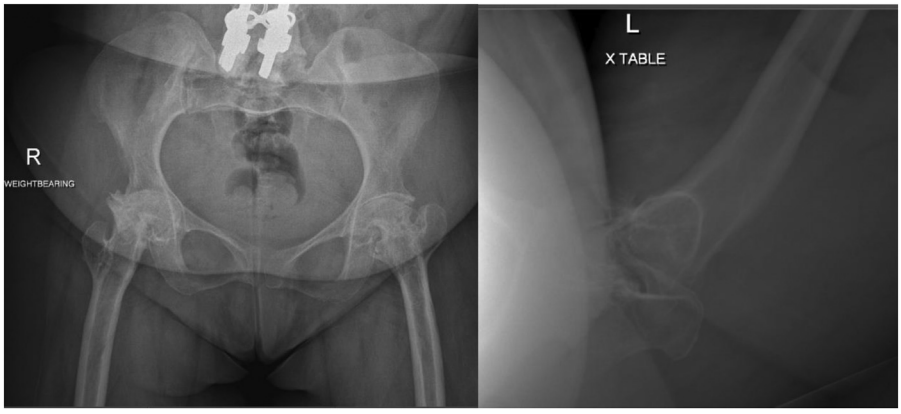


**Figure 1.** Preoperative standing anterior-posterior pelvis radiograph of the pelvis (Left) and cross-table lateral radiograph of the left hip (right).

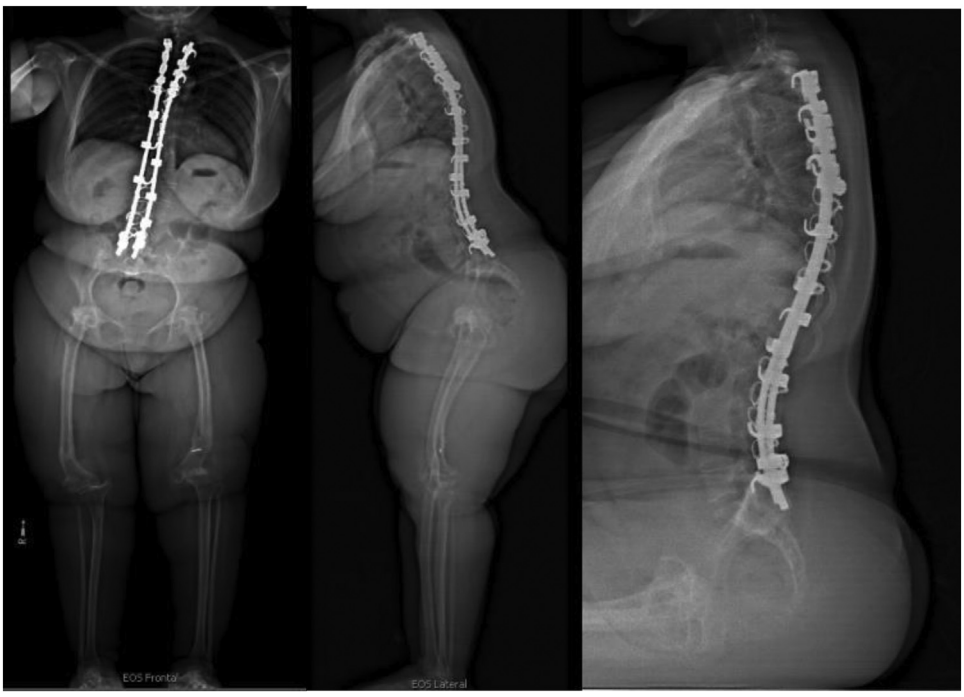


**Figure 2.** Standing full-length anterior-posterior radiograph (Left), standing full-length lateral radiograph (middle), and sitting lateral radiograph (right).

a treatment for end-stage hip arthritis, providers and patients must be educated that this is an option regardless of age. The number of THAs performed annually over the next 10 years is expected to double [17]. This is likely in part a function of an expansion in surgical candidacy, as relatively young age is no longer viewed as an absolute contraindication for surgery. Increased willingness to operate on younger patients is reflected in surgical practice, as database studies have demonstrated the number of THAs performed on patients under 21 years old has substantially increased over the last two decades [18,19].

It is also important to consider that patients with Larsen's syndrome could present with subglottic stenosis, tracheomalacia, unstable cervical spine, odontoid hypoplasia, difficult intubation, and chronic respiratory disease from kyphoscoliosis [20]. If general anesthesia is chosen, it is recommended to obtain preoperative flexion and extension views of the cervical spine, have small tracheal tubes available, and the use of a laryngeal mask airway to minimize manipulation of the neck. When possible, spinal anesthesia is preferred to avoid hyperextension of the cervical spine. It is important that the anesthesia team is aware of the potential problems in patients with Larsen's syndrome and how to manage them, especially in patients with a fused spine, in which spinal anesthesia could be challenging.

Regarding surgical technique, this patient's small stature, osteopenia, and anatomy created several different challenges that are important to recognize and plan for. In addition to proper preoperative imaging studies, it is important to understand the patient's prior surgeries and thus the possibility of distorted surgical anatomy. In this case, the patient had a prior greater trochanteric osteotomy and open hip reductions. This presented a challenge in exposure as well as femoral preparation, as the trochanter was prominent and frail, making it at-risk for fracture.

Acetabular preparation in this case requires planning, as smaller implant sizes are usually required for patients of small stature. Furthermore, using an acetabular component with additional screw-hole options is helpful if obtaining adequate fixation

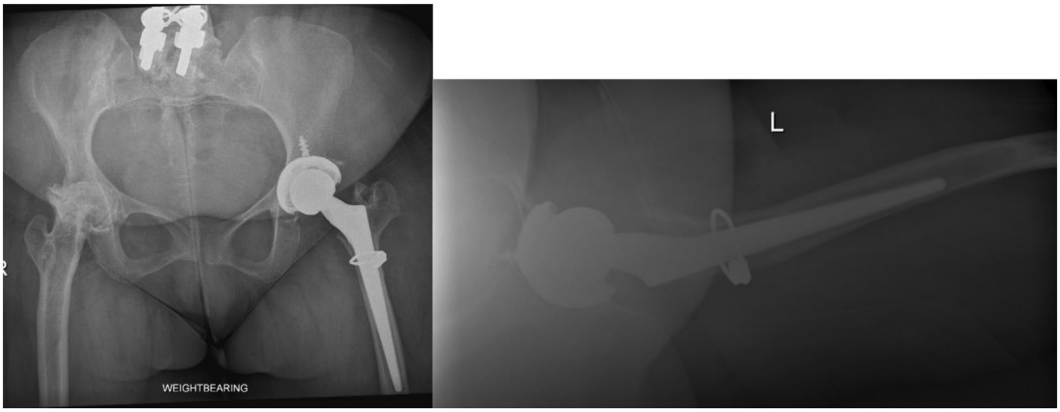


**Figure 3.** Two weeks postoperative left THA.

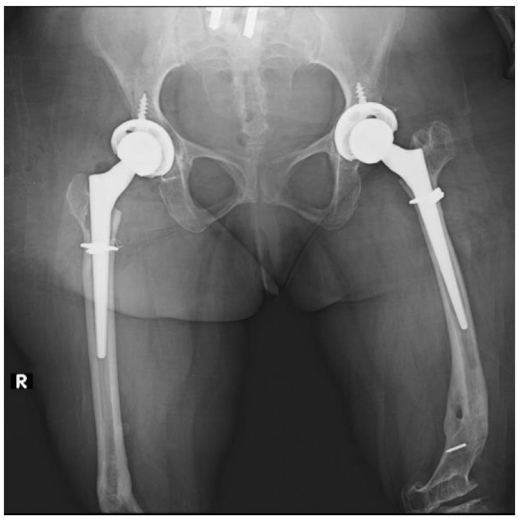


**Figure 4.** Postoperative right THA.

intraoperatively is an issue. If acetabular bone stock is a concern, Judet radiographs or computed tomography should be obtained. If significant protrusion is present, consider an in situ neck cut or other techniques to aid in safe hip dislocation. Navigation could be helpful to assess cup positioning in these cases where traditional anatomic landmarks can be distorted.

This patient has several factors that predispose her to postoperative instability. As is the case with all of our patients, preoperatively, lateral sitting and standing radiographs are obtained to assess spinopelvic mobility. Despite the patient's extended spinal fusion, she has appropriate spinopelvic motion, which we suspect is attributed to the soft tissue laxity associated with her syndrome. The patient's soft tissue quality and tension were noted to be lax intraoperatively. Given the global joint laxity associated with this syndrome, a dual mobility articulation was certainly a consideration; however, due to the patient's small acetabular component size, it was not an option. If a dual mobility construct was an option, it is important to consider the risk-benefit profile of the implant for patients with small sizes. Constructs with smaller inner heads (22 mm and 28 mm) have higher rates of intraprosthetic dissociation events [21,22]. In addition to the added risk of this implant failure, the added stability conferred by dual mobility construct is relatively less in these smaller sizes, and the long-term survivorship of these implants in young patients remains uncertain.

With regards to the femoral component, this patient's proximal femoral geometry posed a challenge. As is the case with small patients with poorly defined canals, care must be taken to safely prepare the femur. In addition to excising soft tissue that may be hindering visualization of the piriformis fossa, it is important to position the leg to optimize trajectory down the axis of the femur. The posterior approach is very effective for this aspect of the case. Next, as identifying and/or reconstituting the femoral canal is challenging, using a burr can be helpful to remove cortical bone that may prevent obtaining a proper start-point for reaming or broaching. The use of intraoperative fluoroscopy or radiographs is encouraged if there is a question during the initial femoral preparation. In the case presented, a short, ream-only, cementless, tapered conical stem was used. This implant obtains fixation below the level of the lesser trochanter through its steep taper and splines and is very useful in cases where proximal fitting is not safe or possible. This implant also allows surgeons to choose anteversion independent of the patient's native proximal femur morphology [23]. Given that the patient's greater trochanters had been previously osteotomized and healed in an anterosuperior position, trochanteric impingement on the ilium and ischium must be considered when choosing the femoral component version. Additionally, placing a prophylactic cable prior to femoral preparation offers protection against hoop stresses and iatrogenic fractures during femoral preparation, especially when using cementless implants.

*Summary*

As is the case with other THA operations, the main goal for THA in this young, low-demand patient is a stable painless hip. Understanding the challenges presented by the patient's anatomy, past surgical history, and the context of her functional status helps guide surgical decision-making and allows the surgeon to perform the procedure safely and give the patient a satisfactory, durable result.

## Conflicts of interest

J.R., N.H., and J.V. have nothing to disclose. R.S. is a paid consultant for Smith & Nephew and Intellijoint. He also has stock options in Gauss Surgical and Pristine Surgical Instruments LLC, outside the submitted work. All authors were not involved in the journal's review of or decisions related to this manuscript.

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## Informed patient consent

The author(s) confirm that written informed consent has been obtained from the involved patient(s) or if appropriate from the parent, guardian, power of attorney of the involved patient(s); and, they have given approval for this information to be published in this case report (series).

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