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### RESEARCH ARTICLE

#### A COST EFFECTIVE TECHNIQUE TOTAL KNEE REPLACEMENT TREATMENT FOR A PATIENT WITH CHARCOT MARRIE TOOTH DISEASE, A CASE REPORT AND LITERATURE REVIEW

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#### Abstract

**Introduction:** Charcot-Marie-Tooth disease affects about 1 in 2,500 individuals. It causes progressive muscle atrophy and sensory loss, predominantly in the distal extremities. CMT manifests early in childhood or adolescence, characterized essentially by features of an unstable gait and a unique type of neurogenic arthropathy known as Charcot joint. Severe muscle weakness and instability is seen in advanced cases of the disease. Charcot joints used to be considered a contraindication for Total Knee Replacement (TKR) due to ligamentous laxity which leads to joint instability. The treatment used to be arthrodesis of the joint and with time rotating stem prosthesis for knee with severe ligamentous laxity or proximal tibia prosthesis in cases of destructive knee joint. In this case we are presenting a rare case which treated by a cost effective technique to overcome the knee joint instability.

**Case Presentation:** We present the case of a 75-year-old female with CMT and multiple co-morbidities. She presented with history of pain in both knees, more in the right knee. She didn't respond to conservative treatment. Her physical examination revealed neuromuscular weakness, tenderness in both knees and restricted range of motion. The X-rays confirmed advanced osteoarthritis without destructive knee joint or subluxation. Preoperatively, the patient underwent physiotherapy to improve range of motion. She was subsequently taken for a right TKR using a semi-constrained system. To overcome the knee instability, a tibia size was 2 with tibia insert size 10 mm and tibia stem 25mm are used and the femur size was 3. Intraoperatively patient showed stable valgus and varus test. Postoperatively, the patient began mobilization and range-of-motion exercises on the first day without instability. Two weeks later, she was ambulating independently with a walker, had significant pain improvement, and nearly full range of motion.

**Conclusion:** The positive outcome in this patient suggests that using a semi-constrained TKR can be successful and cost-effective solution for managing Charcot knee in individuals with CMT, leading to significant improvements in pain and function. This is the first reported use of a

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Semi-constrained TKR instead of a rotating hinge to treat Charcot knee. However, further studies with long-term outcomes are needed to better understand its significance in Charcot knee secondary to CMT.

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### **Introduction:-**

Charcot-Marie-Tooth (CMT) disease is the most common hereditary motor and sensory neuropathy, affecting about 1 in 2,500 [1]. The typical pattern of inheritance is autosomal dominant. It forms a group of disorders characterized by progression of muscle atrophy and sensory loss in different parts of the body. CMT typically presents during adolescence or early adulthood and progresses slowly, causing an unstable gait and a relatively distinctive neurogenic arthropathy recognized as a Charcot joint [2].

The primary effect of the disease is peripheral nerves, causing muscle weakening and atrophy in predominantly distal parts of extremities [3]. The most prominent sign is deformity of the feet, but CMT can cause other conditions such as knee osteoarthritis from the weakening and instability of muscles [4]. Weakened muscular strength, mainly in the lower limbs, may result in abnormal biomechanics during motion and promote joint degeneration, inducing osteoarthritis [5].

The Charcot joints were considered a relative contraindication to total knee replacement previously [6], but the procedure is presently considered appropriate if the underlying disorder is controlled [7]. There have been very few accounts on the treatment of Charcot joints in the knees of patients with CMT.

### **Case Presentation**

This is a 75-year-old female with a history of Charcot-Marie-Tooth, Type 2 Diabetes Mellitus, hypertension, and dyslipidemia. Patient presented with history of pain in both knees, more in the right knee, which was progressive in nature and was affecting her daily activities. She did not respond to conservative treatment, which included pain medication, physiotherapy, intraarticular injections, and special support for her feet. Certain shoes were also prescribed to aid her in ambulation. The patient was seen by neurologist outside of our hospital. as per the report of the neurologist that patient was diagnosed with the CMT. The diagnoses confirmed by history, physical examination and nerve conduction study. However, the neurologist did not specify whether it's type one or type two. Also, the nerve conduction study showed general reduction of distal motor latency, motor, sensory nerve conduction velocity.

On physical examination, the patient had neuromuscular weakness. Her feet showed high arches with preservation of the arches; she also had bilateral knee tenderness to palpation, reduced range of motion, with varus deformity with fixed flexion contraction in both knees, 50 degrees in the right knee and 20 degrees in the left knee. Varus stress test was positive in both knees. X-rays showed advanced osteoarthritis with varus deformity in both knees. Before operating on the patient, we referred her in our neurology clinic, the Neurologist confirmed her diagnosis based on the history, family history of CMT and the physical examination which showed generalized decrease the muscle strength more on the lower limb, decrease sensation and loss of the knee-ankle jerks reflexes.

Total knee replacement surgery was planned. Preoperatively, the patient was prepared with a course of physiotherapy to help her regain her range of motion. At the follow-up visit, she walked into the clinic without support, and it was considered appropriate to proceed with total knee replacement surgery under precaution. A semi-constrained right total knee replacement system was used through a medial parapatellar approach. A femur size 3, tibia size 2 were used with tibial insert of size 10, along with a 25 mm tibial stem for added stability. After that, stability of the right knee examined, it showed good stability on valgus and varus stress test. The surgery was carried out under general anesthesia without a tourniquet or non-depolarizing neuromuscular-blocking drugs. No intraoperative complications were noted and bone quality was fine. After the surgery, physical examination showed a stable knee with neither varus nor valgus deformities. The patient began mobilization and range of motion exercises on the first postoperative day.

Two weeks later, the patient was seen in the clinic. She was ambulating independently with a walker, her pain had improved dramatically, and she had achieved almost full range of motion. An X-ray of the knee was taken and showed an intact prosthesis with good alignment.

**Discussion:-**

On reviewing the literature, we found several case reports for patients with Charcot knee undergoing total knee replacement, but only few of these cases were secondary to CMT. All these cases of Charcot knee secondary to CMT used a rotating hinge knee replacement [8,9]. This is the first case to use a Semi constrained TKR for Charcot knee secondary to CMT, highlighting its novelty.

This 75-year-old female patient is known to have CMT and several comorbidities, moreover she has progressively worsened bilateral knee pain that significantly affects her daily activities. Clinical examination revealed neuromuscular weakness, high arches of the feet, tenderness of both knees, bilateral limitation in range of movement, and varus deformity. Radiographic evaluation confirmed advanced osteoarthritis in both knees with associated varus deformity however, there's no severe joint destruction or joint subluxation.

CMT is an inherited neuropathy, involving predominantly peripheral nerves and manifesting with muscle atrophy, weakness, and sensory loss in the distal extremities[1]. Abnormal gait and biomechanics secondary to CMT increase stresses on the knee joints, thereby accelerating the course of osteoarthritis development [5]. This patient's neuromuscular weakness and altered lower-limb mechanics likely played a significant role in the progression of her knee osteoarthritis. Moreover, our patient has several comorbidities themselves that could have influenced the progression of the disease.

The initial management of our patient was conservative, with a program of physiotherapy directed towards improving the lower limb weakness, and the prescription of specialized foot supports, with some improvements. Before physiotherapy the patient could walk but with the need of support; afterward, a little improvement occurred, and she was able to walk without support, after the physiotherapy she had with us in our hospital. Despite such measures, the patient's symptoms persisted, and surgical measures were mandated. Therefore, total knee replacement has been chosen as a definitive measure for alleviating the pain, thus enhancing the quality of life of the patient.

Arthrodesis used to be the primary surgical management for Charcot knee, aimed at providing stability and alleviating pain. This procedure involves the surgical joining of the knee bones, effectively eliminating motion at the joint, which can be beneficial in cases of severe instability [10,11]. Arthrodesis techniques include spring compression fixators, positive pressure fixators, intramedullary locking nails, and bone grafting [12,13,14,15]. Studies generally found that patients were able to walk without pain using a quad cane, enabling them to independently carry out daily activities and even return to work [14,16]. On average, patient was able to return to partial weight bearing after 16.7 weeks and 21.7 weeks to full weight bearing [15]. A case study utilized the American Knee Society Score (AKSS) for knee and function, showing an improvement from 25 to 85 [17]. However, there are some complications when choosing knee arthrodesis for these patients. Firstly, The entry point for intramedullary nails can be a vulnerable area that is prone to fractures, which can lead to femoral shaft fractures [12]. Secondly, fused knees with limited mobility are more susceptible to traumatic injuries [13].

Another surgical approach use for patients with Charcot knee is total knee replacement. Total knee replacement in patients with Charcot-Marie-Tooth disease can be challenging because the outcome of the charcot knees is less favorable compared with the primary total knee replacement results of patients with primary osteoarthritis [18]. The rate of perioperative complications in a Charcot knee, after total knee replacement, is much more than in the standard population, even showing an increased incidence of dislocations, periprosthetic fractures, instability, deep vein thrombosis and infections [19]. This complication rate is far higher than that for arthroplasty in primary osteoarthritis [19]. On the other hand, total knee replacement showed dramatic improvement in reducing the pain patients endure after surgery for Charcot knee [20]. Besides, improvement was also shown in functional outcomes [20,21,22]. Several studies have been conducted which proved that most of the patients with Charcot knee, who failed to walk before the surgery due to their disability, were able to walk. Even those patients who could not walk fully without support were able to walk with slight support [19]. Besides that, in Charcot knee, total knee replacement resulted in improvement in multiple case reports regarding knee alignment [20,21,22], this is required to extend the life of the prosthesis and hence maintain the knee function for a longer period of time [23].

Preoperative knee instability resulting from bony destruction, extreme range of motion and ligamentous laxity has been also observed in patients with Charcot knee which was linked to decreased knee proprioception [19]. Instability was complicated postoperatively as well, a study showed that 7% of knee arthroplasty was complicated by dislocation which was managed by bracing for six months as well as patellar dislocation were lateral release was

done [21]. Longer stem component and use of rotating hinge prostheses was used as well to address ligamentous instability in another study [20]. Similarly, a proximal tibia prostheses was used in a previous case report for a patient with severe destructive subluxated knee joint and severe varus knee[24]. For the current study, the patient does not have subluxated knee joint or severe varus knee joint. Therefore, we decided to use semi-constrained insert has been used in our patient yielding a positive outcome with a cost-effective alternative for the treatment of Charcot knee.

### Conclusion:-

The positive outcome in this patient suggests that, with appropriate precautions, total knee replacement using a semi-constrained with adding larger insert can be a viable and cost-effective option for managing Charcot knee in individuals with CMT, leading to significant improvements in pain and function. Notably, this is the first reported case of using asemi-constrained TKR instead of a rotating hinge or proximal tibia prosthesis in this context, highlighting the cost effective of the approach and not technical demanding. However, further studies with semi-constrained TKR are needed to better understand its significance in Charcot knee secondary to CMT.

### References:-

1. Barreto LC, Oliveira FS, Nunes PS, et al. Epidemiologic Study of Charcot-Marie-Tooth Disease: A Systematic Review. *Neuroepidemiology*. 2016;46(3):157-165. doi:10.1159/000443706
2. Brust JC, Lovelace RE, Devi S. Clinical and electrodiagnostic features of Charcot-Marie-Tooth syndrome. *Acta Neurol Scand Suppl*. 1978;68:1-142.
3. Banchs I, Casasnovas C, Albertí A, et al. Diagnosis of Charcot-Marie-Tooth disease. *J Biomed Biotechnol*. 2009;2009:985415. doi:10.1155/2009/985415
4. Tahmasebi MN, Amjad GG, Kaseb MH, Bashti K. Total Knee Arthroplasty in Severe Unstable Knee: Case-Report and Literature Review. *Arch Bone Jt Surg*. 2017;5(1):58-62.
5. Casasnovas C, Cano LM, Albertí A, Céspedes M, Rigo G. Charcot-Marie-tooth disease. *Foot Ankle Spec*. 2008;1(6):350-354. doi:10.1177/1938640008326247
6. Singh D, Gray J, Laura M, Reilly MM. Charcot neuroarthropathy in patients with Charcot Marie Tooth Disease. *Foot Ankle Surg*. 2021;27(8):865-868. doi:10.1016/j.fas.2020.11.005
7. Yang JH, Yoon JR, Oh CH, Kim TS. Primary total knee arthroplasty using rotating-hinge prosthesis in severely affected knees. *Knee Surg Sports Traumatol Arthrosc*. 2012;20(3):517-523. doi:10.1007/s00167-011-1590-1
8. Ehara Y, Nakano N, Takayama K, et al. Rotating Hinge Knee Arthroplasty for Charcot Joints of the Knees in Patients With Charcot-Marie-Tooth Disease: A Report of Two Cases. *Cureus*. 2024;16(6):e63154. Published 2024 Jun 25. doi:10.7759/cureus.63154
9. Yoshikawa, R., Kitagawa, A., Inokuchi, T., Shima, N., & Hashimoto, Y. (2020). Staged Bilateral Total Knee Arthroplasty in a Patient with Charcot Knees: A Case Report. *Journal of orthopaedic case reports*, 10(2), 9–12. <https://doi.org/10.13107/jocr.2020.v10.i02.1674>
10. Mei F, Li JJ, Lin J, Zhou D, Xing D. Constrained Condylar Prostheses for the Treatment of Charcot Arthropathy: A Case Report and Literature Review. *Orthop Surg*. 2023;15(5):1423-1430. doi:10.1111/os.13686
11. BRASHEAR HR. The value of the intramedullary nail for knee fusion particularly for the Charcot joint. *Am J Surg*. 1954;87(1):63-65. doi:10.1016/0002-9610(54)90042-2
12. Figueiredo A, Ferreira R, Alegre C, Fonseca F. Charcot osteoarthropathy of the knee secondary to neurosyphilis: a rare condition managed by a challenging arthrodesis. *BMJ Case Rep*. 2018;2018:bcr2018225337. Published 2018 Aug 20. doi:10.1136/bcr-2018-225337
13. HOLT EP. Fusion of the charcot knee. *South Med J*. 1957;50(10):1215-1220. doi:10.1097/00007611-195710000-00002
14. Rebelo T, Morais J, Agostinho F, Abrantes A, Simões N, Simões I. Knee Arthrodesis in a Patient with Charcot Neuroarthropathy Secondary to Familial Amyloid Polyneuropathy: A Case Report. *JBJS Case Connect*. 2017;7(4):e97. doi:10.2106/JBJS.CC.17.00110
15. Drennan DB, Fahey JJ, Maylahn DJ. Important factors in achieving arthrodesis of the Charcot knee. *J Bone Joint Surg Am*. 1971;53(6):1180-1193.
16. Fullerton BD, Browngohl LA. Total knee arthroplasty in a patient with bilateral Charcot knees. *Arch Phys Med Rehabil*. 1997;78(7):780-782. doi:10.1016/s0003-9993(97)90090-3
17. Nozaka, K., Miyakoshi, N., Yuasa, Y., Mita, M., & Shimada, Y. (2019). Simultaneous Total Knee Arthroplasty and Ankle Arthrodesis for Charcot Neuroarthropathy. *Case reports in orthopedics*, 2019, 6136409. <https://doi.org/10.1155/2019/6136409>

18. Tibbo ME, Chalmers BP, Berry DJ, Pagnano MW, Lewallen DG, Abdel MP. Primary Total Knee Arthroplasty in Patients With Neuropathic (Charcot) Arthropathy: Contemporary Results [published correction appears in J Arthroplasty. 2019 Jan;34(1):191. doi: 10.1016/j.arth.2018.09.009]. J Arthroplasty. 2018;33(9):2815-2820. doi:10.1016/j.arth.2018.04.003
19. Babazadeh S, Stoney JD, Lim K, Choong PF. Arthroplasty of a Charcot knee. Orthop Rev (Pavia). 2010;2(2):e17. doi:10.4081/or.2010.e17
20. Parvizi J, Marrs J, Morrey BF. Total knee arthroplasty for neuropathic (Charcot) joints. Clin OrthopRelat Res. 2003;(416):145-150. doi:10.1097/01.blo.0000081937.75404.ed
21. Kim YH, Kim JS, Oh SW. Total knee arthroplasty in neuropathic arthropathy. J Bone Joint Surg Br. 2002;84(2):216-219. doi:10.1302/0301-620x.84b2.12312
22. Soudry M, Binazzi R, Johanson NA, Bullough PG, Insall JN. Total knee arthroplasty in Charcot and Charcot-like joints. Clin OrthopRelat Res. 1986;(208):199-204.
23. Siston RA, Giori NJ, Goodman SB, Delp SL. Surgical navigation for total knee arthroplasty: a perspective. J Biomech. 2007;40(4):728-735. doi:10.1016/j.jbiomech.2007.01.006
24. Alshaygy I, Alzahim M, Alhamdan H, Aljassir F. Total knee arthroplasty with megaprosthesis for idiopathic charcot knee. JAAOS Global Research & Reviews. 2021 Mar 1;5(3):e20.

**Appendix:-****Figure 1:-** Preoperative X-rays showing advanced osteoarthritis with varus deformity.

**Figure 2:-** Postoperative X-rays showing intact prosthesis with no signs of malalignment, loosening or periprosthetic fractures.



**Figure 3:-** Follow-up X-rays showing intact prosthesis with no signs of malalignment, loosening or periprosthetic fractures.

