

World Journal of *Clinical Cases*

World J Clin Cases 2022 August 26; 10(24): 8432-8807



EDITORIAL

- 8432 Evolution of *World Journal of Clinical Cases* over the past 5 years
Muthu S

OPINION REVIEW

- 8436 NF- κ B: A novel therapeutic pathway for gastroesophageal reflux disease?
Zhang ML, Ran LQ, Wu MJ, Jia QC, Qin ZM, Peng YG

MINIREVIEWS

- 8443 Obligate aerobic, gram-positive, weak acid-fast, nonmotile bacilli, *Tsukamurella tyrosinosolvans*: Minireview of a rare opportunistic pathogen
Usuda D, Tanaka R, Suzuki M, Shimozawa S, Takano H, Hotchi Y, Tokunaga S, Osugi I, Katou R, Ito S, Mishima K, Kondo A, Mizuno K, Takami H, Komatsu T, Oba J, Nomura T, Sugita M
- 8450 Diffusion tensor imaging pipeline measures of cerebral white matter integrity: An overview of recent advances and prospects
Safri AA, Nassir CMNCM, Iman IN, Mohd Taib NH, Achuthan A, Mustapha M
- 8463 Graft choices for anterolateral ligament knee reconstruction surgery: Current concepts
Chalidis B, Pitsilos C, Kitridis D, Givissis P
- 8474 Overview of the anterolateral complex of the knee
Garcia-Mansilla I, Zicaro JP, Martinez EF, Astoul J, Yacuzzi C, Costa-Paz M
- 8482 Complication of lengthening and the role of post-operative care, physical and psychological rehabilitation among fibula hemimelia
Salimi M, Sarallah R, Javanshir S, Mirghaderi SP, Salimi A, Khanzadeh S

ORIGINAL ARTICLE**Clinical and Translational Research**

- 8490 Pyroptosis-related genes play a significant role in the prognosis of gastric cancer
Guan SH, Wang XY, Shang P, Du QC, Li MZ, Xing X, Yan B

Retrospective Study

- 8506 Effects of propofol combined with lidocaine on hemodynamics, serum adrenocorticotrophic hormone, interleukin-6, and cortisol in children
Shi S, Gan L, Jin CN, Liu RF
- 8514 Correlation analysis of national elite Chinese male table tennis players' shoulder proprioception and muscle strength
Shang XD, Zhang EM, Chen ZL, Zhang L, Qian JH

- 8525** Clinical value of contrast-enhanced ultrasound in early diagnosis of small hepatocellular carcinoma (≤ 2 cm)

Mei Q, Yu M, Chen Q

- 8535** Identification of predictive factors for post-transarterial chemoembolization liver failure in hepatocellular carcinoma patients: A retrospective study

Yuan M, Chen TY, Chen XR, Lu YF, Shi J, Zhang WS, Ye C, Tang BZ, Yang ZG

- 8547** Clinical significance of half-hepatic blood flow occlusion technology in patients with hepatocellular carcinoma with cirrhosis

Liu D, Fang JM, Chen XQ

- 8556** Which octogenarian patients are at higher risk after cholecystectomy for symptomatic gallstone disease? A single center cohort study

D'Acapito F, Solaini L, Di Pietrantonio D, Tauceri F, Mirarchi MT, Antelmi E, Flamini F, Amato A, Framarini M, Ercolani G

Clinical Trials Study

- 8568** Computed tomography combined with gastroscopy for assessment of pancreatic segmental portal hypertension

Wang YL, Zhang HW, Lin F

Observational Study

- 8578** Psychological needs of parents of children with complicated congenital heart disease after admitting to pediatric intensive care unit: A questionnaire study

Zhu JH, Jin CD, Tang XM

Prospective Study

- 8587** Quantitative differentiation of malignant and benign thyroid nodules with multi-parameter diffusion-weighted imaging

Zhu X, Wang J, Wang YC, Zhu ZF, Tang J, Wen XW, Fang Y, Han J

Randomized Controlled Trial

- 8599** Application of unified protocol as a transdiagnostic treatment for emotional disorders during COVID-19: An internet-delivered randomized controlled trial

Yan K, Yusufi MH, Nazari N

- 8615** High-flow nasal cannula oxygen therapy during anesthesia recovery for older orthopedic surgery patients: A prospective randomized controlled trial

Li XN, Zhou CC, Lin ZQ, Jia B, Li XY, Zhao GF, Ye F

SYSTEMATIC REVIEWS

- 8625** Assessment tools for differential diagnosis of neglect: Focusing on egocentric neglect and allocentric neglect

Lee SH, Lim BC, Jeong CY, Kim JH, Jang WH

CASE REPORT

- 8634** Exome analysis for Cronkhite-Canada syndrome: A case report
Li ZD, Rong L, He YJ, Ji YZ, Li X, Song FZ, Li XA
- 8641** Discrepancy between non-invasive prenatal testing result and fetal karyotype caused by rare confined placental mosaicism: A case report
Li Z, Lai GR
- 8648** Paroxysmal speech disorder as the initial symptom in a young adult with anti-N-methyl-D-aspartate receptor encephalitis: A case report
Hu CC, Pan XL, Zhang MX, Chen HF
- 8656** Anesthetics management of a renal angiomyolipoma using pulse pressure variation and non-invasive cardiac output monitoring: A case report
Jeon WJ, Shin WJ, Yoon YJ, Park CW, Shim JH, Cho SY
- 8662** Traumatic giant cell tumor of rib: A case report
Chen YS, Kao HW, Huang HY, Huang TW
- 8667** Analysis of two naval pilots' ejection injuries: Two case reports
Zeng J, Liu XP, Yi JC, Lu X, Liu DD, Jiang YQ, Liu YB, Tian JQ
- 8673** Beware of the DeBakey type I aortic dissection hidden by ischemic stroke: Two case reports
Chen SQ, Luo WL, Liu W, Wang LZ
- 8679** Unilateral lichen planus with Blaschko line distribution: A case report
Dong S, Zhu WJ, Xu M, Zhao XQ, Mou Y
- 8686** Clinical features and progress of ischemic gastritis with high fatalities: Seven case reports
Shionoya K, Sasaki A, Moriya H, Kimura K, Nishino T, Kubota J, Sumida C, Tasaki J, Ichita C, Makazu M, Masuda S, Koizumi K, Kawachi J, Tsukiyama T, Kako M
- 8695** Retinoblastoma in an older child with secondary glaucoma as the first clinical presenting symptom: A case report
Zhang Y, Tang L
- 8703** Recurrent herpes zoster in a rheumatoid arthritis patient treated with tofacitinib: A case report and review of the literature
Lin QX, Meng HJ, Pang YY, Qu Y
- 8709** Intra-abdominal ectopic bronchogenic cyst with a mucinous neoplasm harboring a *GNAS* mutation: A case report
Murakami T, Shimizu H, Yamazaki K, Nojima H, Usui A, Kosugi C, Shuto K, Obi S, Sato T, Yamazaki M, Koda K
- 8718** Effects of intravascular photobiomodulation on motor deficits and brain perfusion images in intractable myasthenia gravis: A case report
Lan CH, Wu YC, Chiang CC, Chang ST

- 8728** Spontaneous acute epidural hematoma secondary to skull and dural metastasis of hepatocellular carcinoma: A case report
Lv GZ, Li GC, Tang WT, Zhou D, Yang Y
- 8735** Malignant melanotic nerve sheath tumors in the spinal canal of psammomatous and non-psammomatous type: Two case reports
Yeom JA, Song YS, Lee IS, Han IH, Choi KU
- 8742** When should endovascular gastrointestinal anastomosis transection Glissonean pedicle not be used in hepatectomy? A case report
Zhao J, Dang YL
- 8749** VARS2 gene mutation leading to overall developmental delay in a child with epilepsy: A case report
Wu XH, Lin SZ, Zhou YQ, Wang WQ, Li JY, Chen QD
- 8755** Junctional bradycardia in a patient with COVID-19: A case report
Aedh AI
- 8761** Application of 3 dimension-printed injection-molded polyether ether ketone lunate prosthesis in the treatment of stage III Kienböck's disease: A case report
Yuan CS, Tang Y, Xie HQ, Liang TT, Li HT, Tang KL
- 8768** High scored thyroid storm after stomach cancer perforation: A case report
Baik SM, Pae Y, Lee JM
- 8775** Cholecystitis-an uncommon complication following thoracic duct embolization for chylothorax: A case report
Dung LV, Hien MM, Tra My TT, Luu DT, Linh LT, Duc NM
- 8782** Endometrial squamous cell carcinoma originating from the cervix: A case report
Shu XY, Dai Z, Zhang S, Yang HX, Bi H
- 8788** Type 2 autoimmune pancreatitis associated with severe ulcerative colitis: Three case reports
Ghali M, Bensted K, Williams DB, Ghaly S
- 8797** Diffuse uterine leiomyomatosis: A case report and review of literature
Ren HM, Wang QZ, Wang JN, Hong GJ, Zhou S, Zhu JY, Li SJ

LETTER TO THE EDITOR

- 8805** Comment on "Posterior reversible encephalopathy syndrome in a patient with metastatic breast cancer: A case report"
Kunić S, Ibrahimagić OĆ, Kojić B, Džananović D

ABOUT COVER

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The *WJCC* is now abstracted and indexed in Science Citation Index Expanded (SCIE, also known as SciSearch®), Journal Citation Reports/Science Edition, Current Contents®/Clinical Medicine, PubMed, PubMed Central, Scopus, Reference Citation Analysis, China National Knowledge Infrastructure, China Science and Technology Journal Database, and Superstar Journals Database. The 2022 Edition of Journal Citation Reports® cites the 2021 impact factor (IF) for *WJCC* as 1.534; IF without journal self cites: 1.491; 5-year IF: 1.599; Journal Citation Indicator: 0.28; Ranking: 135 among 172 journals in medicine, general and internal; and Quartile category: Q4. The *WJCC*'s CiteScore for 2021 is 1.2 and Scopus CiteScore rank 2021: General Medicine is 443/826.

RESPONSIBLE EDITORS FOR THIS ISSUE

Production Editor: *Ying-Yi Yuan*; Production Department Director: *Xu Guo*; Editorial Office Director: *Jin-Lei Wang*.

NAME OF JOURNAL

World Journal of Clinical Cases

ISSN

ISSN 2307-8960 (online)

LAUNCH DATE

April 16, 2013

FREQUENCY

Thrice Monthly

EDITORS-IN-CHIEF

Bao-Gan Peng, Jerzy Tadeusz Chudek, George Kontogeorgos, Maurizio Serati, Ja Hyeon Ku

EDITORIAL BOARD MEMBERS

<https://www.wjgnet.com/2307-8960/editorialboard.htm>

PUBLICATION DATE

August 26, 2022

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INSTRUCTIONS TO AUTHORS

<https://www.wjgnet.com/bpg/gerinfo/204>

GUIDELINES FOR ETHICS DOCUMENTS

<https://www.wjgnet.com/bpg/GerInfo/287>

GUIDELINES FOR NON-NATIVE SPEAKERS OF ENGLISH

<https://www.wjgnet.com/bpg/gerinfo/240>

PUBLICATION ETHICS

<https://www.wjgnet.com/bpg/GerInfo/288>

PUBLICATION MISCONDUCT

<https://www.wjgnet.com/bpg/gerinfo/208>

ARTICLE PROCESSING CHARGE

<https://www.wjgnet.com/bpg/gerinfo/242>

STEPS FOR SUBMITTING MANUSCRIPTS

<https://www.wjgnet.com/bpg/GerInfo/239>

ONLINE SUBMISSION

<https://www.f6publishing.com>

Complication of lengthening and the role of post-operative care, physical and psychological rehabilitation among fibula hemimelia

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Specialty type: Orthopedics

Provenance and peer review:

Invited article; Externally peer reviewed.

Peer-review model: Single blind

Peer-review report's scientific quality classification

Grade A (Excellent): 0

Grade B (Very good): B, B

Grade C (Good): 0

Grade D (Fair): 0

Grade E (Poor): 0

P-Reviewer: Byeon H, South Korea; Velázquez-Saornil J, Spain

Received: March 23, 2022

Peer-review started: March 23, 2022

First decision: April 13, 2022

Revised: April 27, 2022

Accepted: July 25, 2022

Article in press: July 25, 2022

Published online: August 26, 2022



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Abstract

There is a clear clinical need for efficient physiotherapy and rehabilitation programs during and after bone lengthening and reconstruction for gaining the optimal effect and also prevention or treatment of lengthening side effects. Pin tract infection is the most prevalent side effect during lengthening which could be prevented and treated initially *via* proper wound care. Muscle contractures are typically a consequence of the generated tension on the distracted muscle. It can be managed by physiotherapy initially and surgically in later severe stages. Furthermore, it is essential to avoid muscle contracture development, which is the demonstration of the imbalanced muscle appeals on the joint to inhibit the following subluxation. The knee is the furthestmost affected joint by the aforementioned problem due to the inherent lack of ligamentous and bony stability. Joint stiffness is the other possible unfavorable effect of lengthening. It happens because of extensive muscle contractures or may possibly be attributed to rigidity of the joint following the amplified pressure on the joint surface during the process of lengthening. Physiotherapy and occupational therapy including endurance and strength exercise as well as stretching play an important role during the rehabilitation periods for the prevention and also the treatment of muscle contracture and the following deformity and also joint stiffness. Likewise,

the effect of mental and physical rehabilitation programs should not be overlooked.

Key Words: Fibular hemimelia; Rehabilitation; Reconstruction; Lengthening

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Core Tip: Difficulties that occur during limb lengthening are not avoidable. Pin tract infections are considered the most common adverse effect of lengthening which should be prevented through prophylaxis and treated as soon as possible when occurred. Muscle contractures are usually a result of the tension generated on the muscle due to distraction. It can be managed by physiotherapy initially and surgically in later severe stages. Subluxation or dislocation during lengthening may have been attributed to the preexisting joint instability or imbalanced muscle tension that developed during lengthening. It often happens in the knee joint posteriorly.

Citation: Salimi M, Sarallah R, Javanshir S, Mirghaderi SP, Salimi A, Khanzadeh S. Complication of lengthening and the role of post-operative care, physical and psychological rehabilitation among fibula hemimelia. *World J Clin Cases* 2022; 10(24): 8482-8489

URL: <https://www.wjgnet.com/2307-8960/full/v10/i24/8482.htm>

DOI: <https://dx.doi.org/10.12998/wjcc.v10.i24.8482>

INTRODUCTION

Fibular hemimelia (FH) is considered the most common long bones deficiency and includes an extensive spectrum of anomalies, from a slight partial fibular shortening to a whole lack of fibula, and associated defects of the foot, and ankle, tibia, femur, and even hand abnormality[1].

With respect to the numerous deformities among fibular insufficiency as well as its relationship with postaxial hypoplasia, decision-making and strategies of treatment must be personalized[2]. Treatment choices consist of non-surgical management with shoe lift and orthotic devices; limb preservation, which includes limb elongation strategies; or amputation followed by lifetime prosthetic use. The aforementioned recommended managements are principally hooked on the harshness of the amount of limb length discrepancy, the foot deformity and deficiency, and the potential for achieving the functional and plantigrade foot[3]. The optimal treatment of FH has been controversial for a long time. Amputation along with prosthetic fitting has been recommended as an effective method in the management of FH in early studies and they also use to believe that patients may be more satisfied with amputation and prosthesis with no need for multiple surgeries to correct the deformities and then lengthen the usual major limb length discrepancies[4].

Unfavorable side effects have inundated limb-lengthening procedures since the first introduction for lengthening of the lower limb in 1905[5]. High obstacle rates, mostly bone-healing related, turn into the hallmark of the conservatively accepted aforementioned technique[6]. Along with the introduction of physiological-based approaches to elongation established by Ilizarov[7] in 1988 and according to the biology and physiology of soft tissue as well as bone regeneration under the tension and stretching condition, bone healing problems and complications have developed substantially less and the treatment goals are mostly achieved[8].

It did not take long before the great revolution of Ilizarov's principles of external fixation, became the main pillar of reconstruction and lengthening. This improved the results of limb preservation and reconstruction presenting an attractive alternative to amputation[9]. Over time and as surgeons become more interested in limb preservation, lengthening, and reconstruction, several studies have been conducted on the effects of post-operative care and rehabilitation on the final outcome of reconstructive surgical approaches[10,11]. Since the Ilizarov technique is slow and gradual through daily physiologic changes in the soft tissues and bone, many potential difficulties develop during the distraction as well as fixation periods. Some of the aforementioned difficulties could be solved during the lengthening procedure[6]. In this perusal, we reviewed the literature to find the current post-operative care and rehabilitation programs during and after limb lengthening and reconstruction.

PIN TRACT INFECTION

Pin tract infections are considered the most common adverse effect of lengthening or even an almost inevitable obstacle in using external apparatus with various rates of 5% to 100% which is the result of

pin tract insertion[12,13] (Figure 1). It happens because the natural barrier, the skin, and residues open, providing a communication, vehiculated through the pins, between the bone and the surroundings. It can ultimately cause osteomyelitis, loosening, and fixation loss[14]. Moreover, rehabilitation may lead to motion at the pin-skin interface, aggregating the risk for pin tract infection[15].

Treatment and prophylaxis of pin tract infection are influenced by different parameters[14]. Pins and holes care includes gentle softening and massage of the crust by moistening it with saline, following the use of alcohol, and the application of an antiseptic which usually consists of chlorhexidine or povidone-iodine[16,17]. Some other authors have also used tribromophenate, paraffin ointment, bismuth, silver sulfadiazine, polyhexamethylene biguanide, or hydrogen peroxide[18-21]. Protection with dry gauze finalizes the care[14]. Dry nailing makes the environment of the pinning site unfavorable for germs to grow. The frequency of the aforementioned care is usually every two days, even though it varies between once a week and twice a day[14].

It has been suggested that the prophylactic prudence including the release of the skin tension at the pin-site as well as cautious pin-site overhaul and care by the parents will help in the prevention and control of the infections and their development to a complication[3,22-26]. Likewise, including the knee in the Ilizarov frame prevent moving the proximal ring to the cutaneous or subcutaneous tissue of the posterior thigh or popliteal fossa and further wound infections[27].

MUSCLE CONTRACTURE

Muscle contracture is frequently a consequence of the over tension on the muscle following distraction [28]. They often have a tendency to happen in the over-powering muscle group. It goes back to the strength difference between extensors and flexors[29]. Knee flexion and ankle plantarflexion contracture during the lengthening of the tibia, the most common site of lengthening among fibula hemimelia, occurs because of the triceps surae muscle's resistance to lengthening because of their large muscle mass and strength[30]. In some cases, who have a simultaneous proximal focal deficiency, which is accompanied by fibular hemimelia in 50% to 80%, the second stage of lengthening from femoral bone is desirable[31]. In these cases, the hamstrings are the bulkiest and largest muscle group. The hamstring contracture leads to the moving back of the leg (hip extension) and the knee to bend (knee flexion)[32].

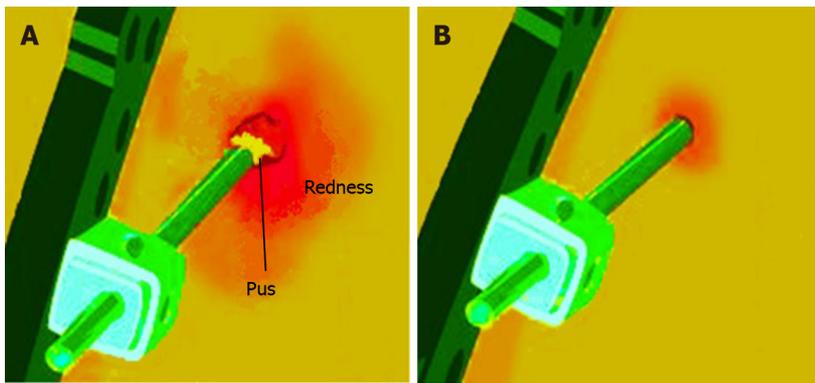
The muscles that cross two joints are frequently involved in contractures because they contain fibers of numerous extents as opposed to muscles crossing a single joint and have fibers of the same extent[33] (Figure 2). The aforementioned statement would cause tension deviations in similar muscles. Muscle tension is a mechanism of stimulating regeneration in muscle during the lengthening procedure. Muscles that pass through two joints may contain a variance proportion of histogenesis in comparison to muscles crossing a single joint[34]. Moreover, it may be attributed to the differences in the maximum histogenesis rate and potential between bone and muscle[35].

The other etiologic concern is the muscles or tendons' transfixion by the pins of the apparatus[36]. It would be amplified *via* transfixed pins, the longitudinal clump of numerous pins in the single plane as well as the thick pins' diameter. Transfixed fascia and tendons lead to joint motion restriction more than transfixed muscles[36].

Therefore, muscle contracture prevention is an inevitable portion of the elongation process. The initial preventive methods include fixation across joints, splinting, and physiotherapy[37]. Physiotherapy has been expected to motivate passive stretching movements for the most involved muscles. Moreover, it is not enough to stretch the muscle only at one end, due to the fact the aforementioned muscle typically crosses two joints. For instance, regarding the triceps surae, while the knee is flexed, the foot should be motivated to dorsiflex maximally. Afterward, the knee would be extended passively, as the ankle is dorsiflexed[38,39]. The patient must be instructed and motivated to repeat the aforementioned movements during the day[40]. Electrical stimulation, as well as active exercises, are approved to improve muscle regeneration[41]. There are several reports which have shown the positive role of continuous passive motions (CPM). Many studies have demonstrated the encouraging effect of passive motion in improving the knee flexion contracture[42,43]. CPM has also some proven hopeful effects in plantar flexion contracture of the ankle which is frequently seen following tibia lengthening[44].

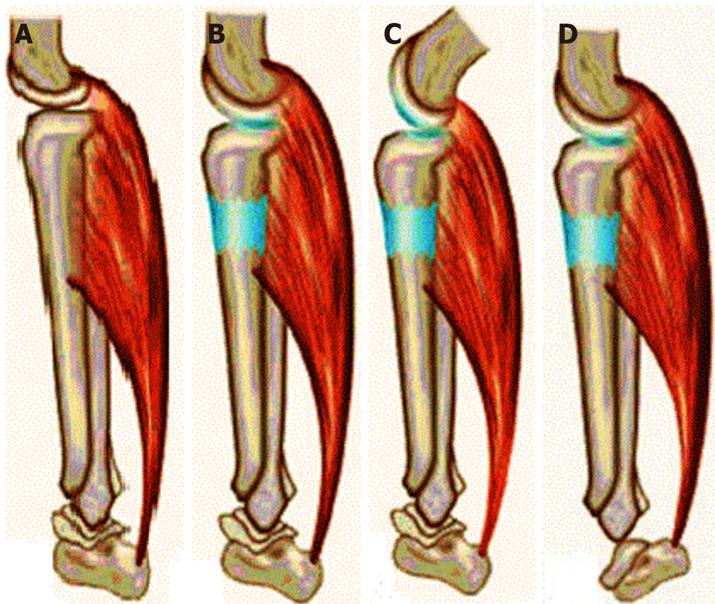
The principles of contracture prevention would work if they have been maintained for at least six hours each day. It is far from the mind that most cases would exercise their muscles that much[45,46].

Trigger point dry needling is suggestive management that would reduce the pain during the physiotherapy and stretching process. This hypothesis has been supported by several investigations demonstrating that muscle damage identified by magnetic resonance imaging after the application of dry needling over the spasmed muscle was associated with post-needling induced pain[42]. They have associated the presence of post-needling with the number of local twitch responses elicited during the needling therapy. Moreover, Gattie *et al*[47] conducted an interesting meta-analysis that displayed low to moderate evidence suggesting that dry needling, when applied by physical therapists, is superior to each treatment solely. This may reduce the pain and so fascinate the stretching progression.



DOI: 10.12998/wjcc.v10.i24.8482 Copyright ©The Author(s) 2022.

Figure 1 Pin tract infection. A: Healthy healing of pin tract site; B: Infection of pin tract site.



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Figure 2 Lateral view of the tibia and associated muscles. A: Before lengthening. Gastrosoleus muscles and Achilles tendon; B: After lengthening of tibia. Ideally the muscle should lengthen to the same extent as the bone; C: Knee flexion contracture. This occurs when knee extension is not maintained throughout lengthening. A relative shortening of the muscle to the new bone length has been produced; D: Equinus contracture of ankle. This occurs when ankle dorsiflexion is not maintained during lengthening.

SUBLUXATION/ DISLOCATION

Dislocation or subluxation of the adjacent joint frequently occurs during lengthening[48]. It may have been attributed to the preexisting joint instability. For example, the anterior cruciate ligament deficiency almost always occurs in the fibula hemimelia state due to femoral notch narrowing[49,50]. The imbalanced muscle pressure that developed during elongation might conduce subluxation, even in the lack of preoperative instability; this particularly happens in the knee[51,52]. Therefore, it is fundamental to avoid muscle contracture presentation.

The knee is the most frequently affected joint because of the inherent deficiency of bony and ligamentous stability[28]. It also should be mentioned that if there is a body segment with a hypomobility (deficit of movement), the nearest segment (joint) will assume that deficit creates hypermobility (increased movement, and therefore the possibility of subluxation) thus creating a compensatory mechanism automatically and involuntarily by the patient.

The subluxated joint may be improved using stretching physiotherapy of the distorting muscle force. It is worth mentioning that physiotherapy would help in the early stages and among milder cases. Traction is another way of management[53]. For the cases of complete dislocation as well as the more severe and persistent subluxation, the apparatus should be extended across the joint to distract and then relocate it gradually or immediately. Moreover, tendon or capsular releases or reapplication of the Ilizarov may be needed[54].

JOINT STIFFNESS

Joint rigidity is usually known as a late-onset complication[55]. This happens because of long-term muscle contractures or can be attributed to rigidity of the joint following the amplified pressure on the joint surface during the elongation process[56]. If a joint is being assumed at the risk of residual rigidity while the elongation device was still on, it should be extended across the joint, and the joint can be distracted afterward. Following that the device can be used to mobilize the joint prior to its removal [57]. The severity of adverse effects can be different according to the created functional limitation. Apparently, 15° loss of ankle dorsiflexion or knee extension is much more significant in comparison to the same motion limitation in ankle plantar flexion and knee flexion[58].

PSYCHOLOGICAL AND MENTAL REHABILITATION

Many experienced pediatric orthopaedics have stated that quality of life will increase as the child grows up[22,59]. The higher quality of life in adults may be attributed to psychological reasons and musculo-skeletal development[60]. To describe more precisely, children and their parents are generally under the impression that surgery will make their appearance, daily function, and other physical aspects normal and like healthy individuals[61]. They sometimes confuse this defect with a simple bone fracture and expect it to be completely repaired with surgery. But as these patients age, they will become more realistic about their malformation and learn to adjust their lives to their disease[22]. Therefore, more involved in routine daily activity and report a higher quality of life when being surveyed. Through much literature review, we have come to the conclusion that educating parents about the fact that FH has a range of problems and that the patients who choose any management method will never be as skeletal as their peers are ultimately effective in their satisfaction[61].

CONCLUSION

Limb salvage and lengthening is a speedily growing approach to orthopaedic surgery among bone deficient cases. There are some unavoidable adverse effects. Pin Tract Infection could be extensively prevented *via* post-operative wound care and cleaning. Muscle contracture could be prevented and managed by physiotherapy in the early stages and surgically in later severe stages. Subluxation/dislocation would be treated by stretching the deforming muscle force during the physiotherapy procedure. Moreover, if a joint is being assumed at the risk of residual rigidity while the elongation device was still on, it should be extended across the joint, and the joint can be distracted afterward. Following that the device can be used to mobilize the joint prior to its removal. Therefore, the aforementioned issues during or after lengthening could be prevented extensively through proper physiotherapy and occupational therapy including endurance and strength exercise as well as stretching programs. Psychological Rehabilitation would also improve the long-term quality of life and patients' satisfaction.

FOOTNOTES

Author contributions: Salimi M performed the majority of the writing; Mirghaderi SP prepared the figures; Salimi A performed data accusation and writing; Sarallah R and Khanzadeh S provided the input in writing the paper; Javanshir S designed the outline and coordinated the writing of the paper.

Conflict-of-interest statement: There is no conflict of interest associated with any of the senior author or other coauthors contributed their efforts in this manuscript.

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Country/Territory of origin: Iran

ORCID number: Maryam Salimi 0000-0001-9771-7048; Amirhossein Salimi 0000-0002-8381-1022.

S-Editor: Yan JP

L-Editor: A

P-Editor: Yan JP

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