

Delayed Prosthesis Fitting in an Elderly with Congenital Lower Limb Deficiency: A Case Report*

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ABSTRACT

Congenital anomalies, including limb deficiency, affect approximately 7.9 million newborns annually. Fitting the prosthesis at an early stage will aid the patient to achieve two-legged standing, to develop reciprocating gait and to attain an optimal body image. A 64-year-old female diagnosed with congenital limb deficiency, right, using a customized shoe was admitted for prosthetic management at the study hospital.

Patient underwent inpatient prosthetic training with physical and occupational therapy. Psychological evaluation was done to assess for depression and anxiety and to determine the patient's response to the prosthesis. The Prosthesis Evaluation Questionnaire (PEQ), an instrument to measure prosthesis-related quality of life and functional outcome, was administered. 3D motion analysis was done to observe the temporospatial parameters of the patient's gait with and without the prosthesis.

The patient had generally positive response towards her prosthesis based on PEQ administered 1 month after the provision of prosthesis. Psychological evaluation showed that the patient had improved general outlook after the provision of the prosthesis. Patient had slower self-selected walking speed compared to transtibial amputee using prosthesis.

Early fitting and fabrication of prosthesis is recommended for the patient with congenital amputation. Studies showed that delayed fitting of prosthesis may decrease acceptance and use of prosthesis. However, provision of properly fitted prosthesis may still improve the quality of life of the patient who is already functionally independent without it. More than increase in physical activity, participation in leisurely activities that promote social interaction motivates elderly patients to use their prosthesis more often.

Key Words *elderly, congenital limb deficiency, leg prosthesis, delayed fitting*

*Second place, Case Report Category, PMA Residents' Research Competition 2016

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INTRODUCTION

Congenital anomalies, including limb deficiency, affect approximately 7.9 million newborns annually. In the Philippines, the burden of birth defects is not yet well established.¹ However, in the Philippine Birth Defects Registry Project of the Department of Health and Human Genetics-National Institutes of Health University of the Philippines Manila, musculo-skeletal congenital malformations ranked 7th among the identified top ten birth defects in 79 hospitals in the country in 1999-2000.² Out of the 450 patients admitted at the amputee ward of the Department of Rehabilitation Medicine (DRM) of the study hospital from 2011 to 2015, 17 patients had congenital limb deficiency. Among the 17, only 4 were adults while the rest were pediatric patients.³

The basic principle in prosthetic management in patients with congenital limb deficiency is to provide the prosthesis, which is appropriate for the developmental stage. It is recommended to start fitting the patient with lower limb prosthesis between the ages of 7 to 14 months. At this time, the patient is almost ready to pull up to stand.⁴ Fitting the prosthesis at this early stage will aid the patient to achieve two-legged standing, to develop reciprocating gait and to provide a complete body image. Even without formal prosthetic gait training, children generally do well with lower limb prosthesis.⁵

Previous studies have been done among elderly amputees to determine the criteria for prescribing prosthesis,⁶ factors that affect the use of the prosthesis,^{7,8} functional outcome after provision of prosthesis,⁹ as well as the level of satisfaction with prosthesis among elderly amputees.¹⁰ However, the reasons for amputation among those included in these studies are vascular diseases, trauma or malignancy.

Among the 18 to 84 years old who have had major upper- or lower-limb loss due to vascular disease, trauma, or malignancy included in the study of Pezzin in 2004, 94.5% had prosthesis and used it extensively (71 hours/week). Seventy six percent appeared to be satisfied with the overall performance of their prostheses. Frequency of prosthesis use and satisfaction with the device were significantly higher among those with shorter timing to first prosthesis fitting. There were no significant differences in prosthesis use, satisfaction, or assessment of prosthetists' quality based on amputation etiology or

amputation level.¹¹ In the study of Spruit-van Eijk et al, elderly patients who underwent prosthetic training have higher probability of using their prosthesis when they are able to ambulate independently after the transtibial amputation.¹² Studies among congenital amputees, on the other hand, included children or the younger age group.^{13,14}

Pezzin et al agreed with results of previous studies that suggest a strong negative association between timing to first prosthesis fitting and use of and satisfaction with the device. When prosthesis fitting is delayed patients seem to be less accepting of the prosthesis and use of prosthesis is also lesser.¹¹ This case report aims to explore the motivation and acceptance of prosthesis of an elderly with congenital amputation.

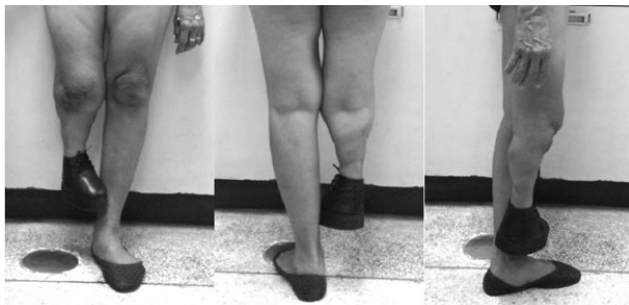
Case Report Proper

A 64 year old single female from Pandacan, Manila consulted at the outpatient clinic of the Department of Rehabilitation Medicine (DRM) of the study hospital last February 13, 2015 with a chief complaint of recurrent callous formation on the distal end of the stump of the right lower extremity. An informed consent was secured from the patient to report this case.

The patient was born with a congenital deformity of the right lower extremity, which is shorter compared to the left and an absent foot. She started to ambulate independently at the age of 2 years old, with weight bearing on the distal end of the right lower extremity. At home, the patient did not use any covering for the distal end of her leg. In the community, however, she would sometimes use a rubber slipper strapped on the stump. The patient was able to finish up to high school. She worked as manicurist and a hair-stylist. Her job entails standing for prolonged hours and sometimes walking long distances to provide service to her clients. At approximately 30 years old she was advised by her client to have a shoe customized to cover the distal end of the stump (Figure 1 and 2).



Figure 1. Customized shoe for the right foot



Figures 2. Patient using her customized shoe on the right foot, (A) anterior, (B) posterior, (C) right lateral views

When the patient was approximately 40 years old (between 1997-1999), the patient consulted a physician in the local health center in Pandacan, Manila, due to the ulceration around a callous on the distal end of her stump. She was referred to a surgeon who in turn advised revision of the stump. However, the patient did not consent for the said procedure. She was referred to Tahanang Walang Hagdan, Inc. (TWHI) for provision of lower limb prosthesis and to Philippine Charity Sweepstakes Office (PCSO) for funding. However, she did not undergo prosthetic training. The patient was able to use the prosthesis for only a month due to improper fit of the socket and heavy weight of the prosthesis. She noted that weight bearing was still on the distal end of the stump. She then returned to using a custom-made shoe for her right lower extremity.

In 2013, the patient experienced aching pain localized over the left knee after prolonged walking or standing. She again consulted in the local health center; impression then was beginning osteoarthritis of the left knee. The physician referred her to the Philippine School of Prosthetics and Orthotics (PSPO) for possible prosthetic management of the right lower extremity to provide a more even weight distribution. Upon assessment, she was prescribed with a transtibial prosthesis: patellar-tendon bearing bi-valve socket with provision to accommodate the stump, exoskeletal shank, and solid ankle cushion heel (SACH) foot (Figure 3). She also underwent prosthetic training on outpatient basis for one month. After the training, the patient was able to ambulate independently using the prosthesis without a gait aid. Patient was then lost to follow up because she noted improvement in her posture and ambulation.

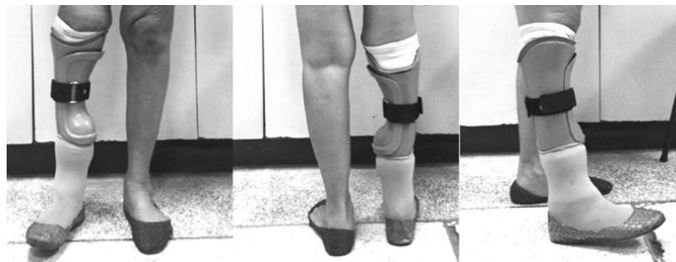


Figure 3. Patient using the prosthesis from PSPO on the right lower extremity, (A) anterior, (B) posterior, (C) right lateral views

However, after one month of using the prosthesis everyday almost the whole day, patient experienced discomfort on the distal end of the stump. The patient could not walk more than 5 steps while using the prosthesis due to severe pain, 8/10 on the numeric pain rating scale, on the distal end of the stump. It was associated with a solitary, well-defined plaque with purulent discharge also on the distal end of the stump. She noted the socket of the prosthesis to be loose resulting to weight bearing on the distal end of the stump. She then stopped using the prosthesis and refrained from bearing weight on the end of the stump until the wound healed.

The patient again used the custom-made shoe during ambulation. However, the ulceration of the callus on her stump would recur. Hence the patient consulted at the out patient clinic of the of the study hospital last February 2015.

Patient's vital signs were essentially normal. Pertinent physical examination findings were: 14-centimeter leg length discrepancy, the right being shorter than the left. True leg length was measured at 85.5 centimeters on the left and 71.5 centimeters on the right. Bilateral femoral length was measured at 50 centimeters. On standing, distal end of the right leg was at the level of the distal third of the left lower leg (Figure 4).



Figure 4. Lower extremities in standing (lateral and anterior views, respectively).

On gross inspection posteriorly while standing on her left leg, shoulder asymmetry was noted, the right was lower than the left but without discrepancy in the muscle bulk. Pelvic obliquity was also noted, the right was also lower than the left. However, there was no noted head tilt towards either side. On sagittal view, no noted excessive kyphosis or lordosis. There was also no noted flexion or extension of the neck. Bilateral hips, knees and left ankle showed full joint range of motion. Left lower extremity had 5/5 muscle strength while the right had 4/5. There were no sensory deficits on both lower extremities. A 2 x 2 centimeter, solitary, well-defined plaque with purulent discharge was noted on the plantar surface of the distal end of the right stump (Figure 5). There were no gross deformities noted on the upper extremities and the left lower extremity. She ambulates independently without any gait aid on level surfaces and negotiates stairs with step over step gait pattern.

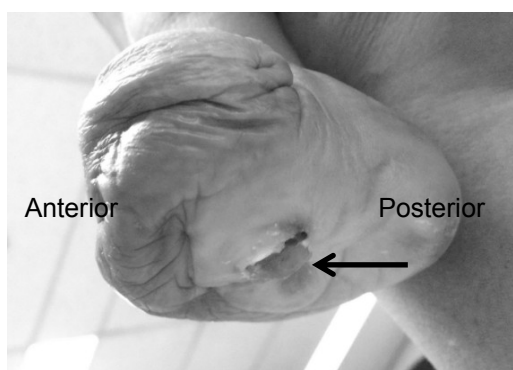


Figure 5. Arrow points to the callus on the plantar surface of the distal end of the stump

She was known to have dyslipidemia and maintained on simvastatin 10mg/tablet once a day. There is no known family history of congenital deformities, hypertension or diabetes. She is not a smoker and not an alcoholic beverage drinker. She lives with her 22-year-old adoptive daughter in a single-storey house with toilet and bath outside the bedroom. She does home-service hair-styling and owns a jeepney for rent.

Radiologic studies done last February 2015 showed widened distal tibia with note of cortical irregularities along the inferior border, which appeared to be the remnant tarsal bones, fused with and exhibited an ill-defined plane of differentiation with the distal tibia (Figure 6). There was also narrowing of the fibular diaphysis with associated tapering of the end.

Overlapping soft tissue density was seen in the distal leg, which may be compatible with the clinically apparent stump. The spine, upper extremities and left lower extremities showed unremarkable radiologic findings. Patient was diagnosed to have transverse tibia partial, fibula partial, tarsus total, metatarsus total, phalanges total, right based on the radiologic findings.

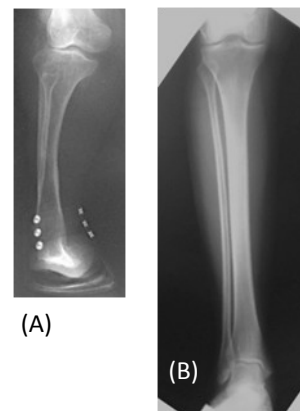


Figure 6. Radiographs of the lower extremities (A: right, B: left)

The following rehabilitation problems were identified: congenital deficiency of the right lower extremity causing a 14-centimeter leg length discrepancy and postural deviation, callus on the plantar aspect of the distal stump and pain over the same area, body image issues, and poor compliance to follow-up. Shown in Figure 7 is the patient's problems using the International Classification of Functioning, Disability and Health (ICF) framework of the patient.

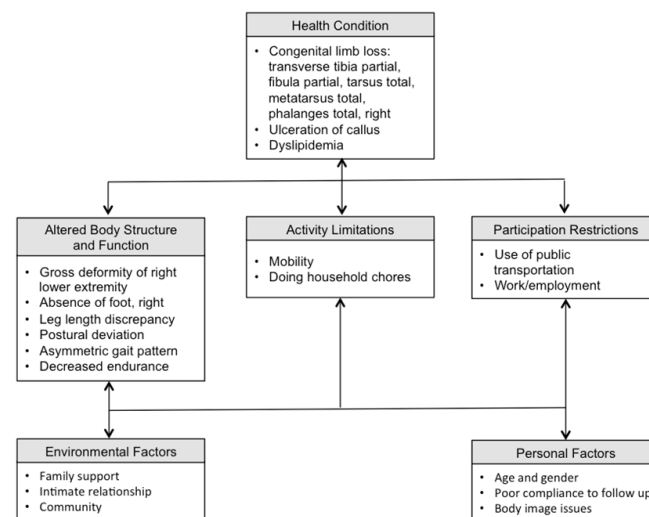


Figure 7. ICF Model

Patient was referred to DRM Psychology Section for psychological assessment prior to the acquisition of a new prosthesis. The initial evaluation indicated that the patient had signs of moderate depression. Although she reported that she has a positive outlook in life, her rating in the Zung's Depression Scale showed that she feels sad most of the time. Using the Taylor's Manifest Anxiety Scale, the patient showed moderate anxiety. She is mostly concerned with her daughter and their finances. Despite the signs of depression and anxiety, the patient was noted to have positive active coping style, high self-esteem, high level of motivation and had realistic expectations. She is confident that she will be able to perform tasks given to her and overcome possible challenges. She however perceived her disability negatively because she thinks persons with disabilities are being pitied on.

The patient was prescribed with below-knee prosthesis with the following components: solid ankle cushion heel (SACH) foot, pylon endoskeletal shank, bi-valve patellar-tendon bearing socket with provision to accommodate shape of the distal stump (Figure 8). This was fabricated at the DRM Prosthetics and Orthotics (P&O) Section.

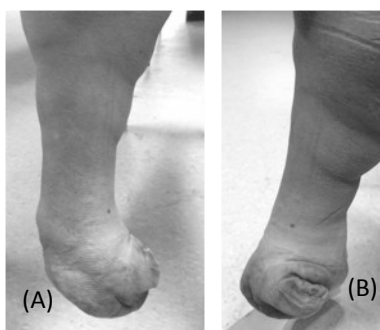


Figure 8. Distal end of the stump, right lower extremity, (A) anterior and (B) medial views.

Prior to the fabrication of the prosthesis, the patient underwent pre-prosthetic training from May to June 2015 at the out patient clinic of the DRM Section of Physical Therapy for 4 weeks, 1 hour/session twice a week. Since the patient had good standing balance and tolerance and was already able to ambulate independently without an assistive device on level surfaces, ramps and stairs. Exercises were focused on mobilization of joints of both upper and lower extremities and the trunk, and endurance training.

While waiting for her prosthesis, the patient went back for follow-up at the PSPO. She reported that she was not able to use the previous prosthesis provided to her because of the improper fit of the socket after one month of daily use. Because she wanted to have 2 prostheses that she can use alternately, she was again prescribed with transtibial prosthesis: patellar-tendon bearing bi-valve socket with provision to accommodate the stump, exoskeletal shank, and solid ankle cushion heel (SACH) foot. Patient however, did not undergo prosthetic training in the said institution.

To facilitate comprehensive rehabilitation management, the patient was admitted at the amputee ward of the study hospital last July 30 to August 7, 2015. Patient was admitted to improve patient compliance to the rehabilitation management, which included physical therapy, occupational therapy, and psychological evaluation and therapy. The goals for admission were as follows: to provide the appropriate prosthesis design; to improve standing balance and tolerance while using the prosthesis, and to facilitate independent ambulation on even surfaces, ramps and stairs with prosthesis; to address body image issues; and to improve compliance with use of prosthesis.

Prosthetic check out was done with the following findings: the prosthesis can stand alone with proper alignment and adequate height, can easily be donned on by the patient, comfortable on standing and sitting with satisfactory antero-posterior and medio-lateral alignment. On standing, there was noted improvement in the shoulder asymmetry and pelvic obliquity previously noted. Patient reported weight bearing on the patellar tendon area of the right lower extremity. No pressure or pain was felt on the distal end of the stump. On walking, there was no noted pistoning or vertical translation of the prosthesis. Shown in Figure 9 is the patient using the prosthesis on anterior, posterior and right lateral views. While using the prosthesis, the patient was independent in transition from sitting to standing but she had fair standing balance and tolerance.



Figure 9. The patient wearing the transtibial patellar tendon bearing prosthesis, (A) anterior, (B) posterior, (C) right lateral views.

She underwent daily 1-hour physical therapy focusing on improving her standing balance and tolerance from fair to good. This was progressed to ambulation training between parallel bars, onto even surfaces outside of the parallel bars, ramps. She was also trained to negotiate stairs using step-by-step gait pattern. Proper posture while standing and walking was also emphasized.

She also underwent daily 1-hour individual, and a weekly group occupational therapy. This focused on retraining in performance of activities of daily living (ADLs) while using prosthesis, proper donning/doffing of prosthesis and lower garments while using the prosthesis, and proper care of the prosthesis.

Daily 1-hour counseling with the psychologist was also provided during her inpatient rehabilitation, which focused on addressing the patient's depression and anxiety. Patient developed a more positive attitude once she received her prosthesis and actively participated in therapy. A visit from her daughter also helped her become more relaxed and thus more compliant to therapy.

To assess for the patient's psychological response to her new prosthesis, she was again referred to psychology for reevaluation after one month of prosthetic use. It was observed that patient's mood has changed since the last assessment. She had a lower score on the Zung's Depression scale. However, she stated that she was still anxious but was now concerned about her daughter more than her prosthesis. Patient valued her independence and ability to achieve her goals. She reported improvement in her mobility and ability to participate in leisurely activities. Being able to walk around more with her daughter and to interact with more neighbors has influenced her satisfaction towards her prosthesis. She also reported positive comments from her clients and neighbors regarding the change in her gait, posture and over-all appearance. The comments, more than the improvements, made her appreciate her prosthesis more.

The Prosthesis Evaluation Questionnaire (PEQ)¹⁵, is a validated questionnaire composed of multiple questions and 9 validated scales which include: ambulation, appearance, frustration, perceived response, residual limb health, social burden, sounds, utility, and well-being. All scales were scored so that 100 indicated the best outcome. The PEQ was administered to the patient after 1 month of using the prosthesis. She had generally positive response

regarding her prosthesis over the past 4 weeks. This time she scored high (greater than 50) on all the scales: ambulation (79.83), appearance (65), frustration (100), residual limb health (83.5), social burden (83), sound (100), utility (60.38), and well being (73) (Appendix B). However, the perceived response scale was still not included because more than half of the questions had "no response" (NR) scores thus making it invalid. Results of the PEQ are summarized in Appendix A.

Last September 11, 2015, a quantitative gait analysis was done using Optitrack System at the Motion Analysis Program of the Department of Orthopedics with the patient using the custom made shoe and using prosthesis. With the patient using the custom-made shoe, the measured speed was 0.55 meters/second, step length was 0.35 ± 0.24 meters on the left and 0.30 ± 0.29 meters on the right with the custom made shoe, and stride length was 0.65 ± 0.19 meters (Table 1).

Table 1. Gait Analysis: using the custom-made shoe done last September 11, 2015

Measure + Std Dev	Left	Right
Step Length (meter)	0.35 ± 0.24	0.30 ± 0.29
Steps/Minute	205.94 ± 18.06	124.36 ± 86.32
Strides/Minute	49.57 ± 1.30	54.04 ± 14.60
Speed (meters/second)	0.55	
Stride Length (meter)	0.65 ± 0.19	

With the patient using the prosthesis, the measured speed was 0.42 meters/second, step length was 0.26 ± 0.04 meters on the left and 0.41 ± 0.05 meters on the right with the prosthesis, and stride length was 0.67 ± 0.08 meters (Table 2).

Table 2. Gait Analysis: with prosthesis done last September 11, 2015, 1 month after prosthetic training

Measure + Std Dev	Left	Right
Step Length (meter)	0.26 ± 0.04	0.41 ± 0.05
Steps/Minute	83.10 ± 5.69	71.06 ± 5.16
Strides/Minute	37.89 ± 1.55	38.58 ± 2.57
Speed (meters/second)	0.42	
Stride Length (meter)	0.67 ± 0.08	

DISCUSSION

In patients with congenital amputation limb deficiency, prosthetic management is more than providing the proper prosthesis for the patient. It is very important to provide prosthetic care that will promote normal sequence of development of the patient. Timing of fitting of prosthesis is important and is usually recommended to start as early as the child pulls up to stand, around the age of 7 months.¹⁴ The psychological impact of the child's deformity on the family should also be addressed. Very young children may not have awareness of their disability but may learn about it through their family's reaction. Because of their disability, people around them tend to treat them differently. According to the patient, she has accepted her disability because her family and close relatives did not treat her differently. She became independent in performing activities of daily living, was able to care for her daughter and was able to work despite her disability. Patient however, grew up avoiding gatherings and occasions because she was preoccupied with other people's opinion of her. She was only able to obtain her first lower limb prosthesis when she was already an adult, at around 40 years old. Almost 15 years later, she again had another prosthesis made despite not being able to use the first one. The patient was noted to have poor compliance to follow up. Whenever she had a problem with her prosthesis, she would just stop using it.

Bilodeau et al studied 65 unilateral vascular amputees 60 years old or over to determine the factors that influence the use of disuse of prosthesis by the elderly amputee.⁷ They explored the influence on the use of prosthesis of the following factors: age, gender, level of physical disability, cognition, self-perceived health and the amputee's satisfaction towards the prosthesis. Elderly amputees may have medical and mental problems that could lead to lower potential for prosthetic management and rehabilitation. They reported that 81% of the subjects wore their prosthesis everyday and 89% among them wore it 6 hours or more per day. Among the factors studied, physical independence had the highest correlation with use of prosthesis. Patients use their prosthesis more often as they become more independent. Similar to other studies, they concluded that age was negatively associated with use of prosthesis. The older patients tend to use their prosthesis or do not use it at all.¹⁶

In contrast to the patient's case, she was already independent prior to using the prosthesis. The patient was already an adult, ~ 40 years old, when she sought prosthetic management. Probably, physical independence was not the motivation for the patient to procure and use her prosthesis. Although the patient was diagnosed to have depression and high level of anxiety, it was reported by Bilodeau et al that depression was not significantly associated with the use of prosthesis.⁷

Legro et al explored the issues of importance reported by persons with lower limb amputations and prostheses using the PEQ. The author concluded that ambulation was the most important function of the prosthesis and the way it fit the residual limb is the most important characteristic of the prosthesis.¹⁵ The responses on PEQ of the patient was similar to the study, wherein she had more positive reaction towards the fit of her prosthesis especially now that the callus is gradually improving.

Murray and Jezz studied the correlation between body image and prosthesis satisfaction in the lower limb amputee. They concluded that body image and prosthesis satisfaction had a close relationship among lower limb prosthesis users.¹⁶ There were also gender variations in this relationship. Functionality influenced the prosthesis satisfaction among male amputees. On the other hand, the aesthetic components influenced the females' prosthesis satisfaction.¹⁶

Scientific evidence has supported that physical activity can prevent and treat disease. It is important in healthy individuals because it promotes a sense of well-being. Among amputees, use of prosthesis can allow or improve physical activity. Deans et al explored the relationship between physical activity and the perceived quality of life among lower limb amputees due to peripheral vascular disease. They observed a relationship between physical activity and the accompanying perception of increased quality of life but there was a weaker-than-expected relationship between the two. They noted however that maintaining social interaction, especially in the elder amputees, had stronger relationship with improved quality of life.¹⁸ These patients are motivated to participate in rehabilitation so long as it can promote and maintain social interaction. This could also be true for the patient. Although she was functionally independent without a lower limb prosthesis, she is highly motivated to acquire and use her prosthesis now so she could participate in leisurely activities with her daughter and her neighbors.

CONCLUSION AND RECOMMENDATION

Early fitting and fabrication of prosthesis is recommended for the patient with congenital amputation. However, some patients may already learn to adapt to their disability and become independent despite the amputation. This influences the level of acceptance of the prosthesis. Studies showed that delayed fitting of prosthesis may decrease acceptance and use of prosthesis. However, provision of properly fit prosthesis may still improve the quality of life of the patient who is already functionally independent without it. More than increase in physical activity, participation in leisurely activities that promote social interaction motivates elderly patients to use their prosthesis more often. The patient will still benefit from a comprehensive rehabilitation management, which includes appropriate and properly fit prosthesis, physical and occupational therapy, and psychotherapy.

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Appendix A: Prosthetic evaluation questionnaire administered 4 weeks after using the prosthesis, administered last September 2, 2015.

Validated Scale Name	Questions	Score	
Ambulation (AM)	13A	80	
79.83	13B	78	
	13C	79	
	13D	82	
	14E	NR	Not tried walking uphill
	14F	NR	Not tried walking downhill
	14G	80	
	14H	80	
Appearance (AP)	3J	48	
65	3M	100	
	3N	NR	Prosthesis has no cover
	4O	0	
	4P	12	
Frustration (FR)	10B	100	
100	10C	100	
Perceived Response (PR)	10A	100	
Not valid since more than half of the items are scored NR.	11D	NR	She has no partner
	11E	NR	She has no partner
	11G	82	
	12H	NR	No other close family member aside from daughter
Residual Limb Health (RL)	4Q	100	
83.5	4R	76	
	4S	100	
	5T	100	
	5U	100	
	5V	25	
Social Burden (SB)	12I	100	
83	12J	100	
	12K	49	
Sounds (SO)	3K	100	
100	3L	100	
Utility (UT)	1B	50	
60.38	1C	100	
	1D	48	
	2E	100	
	2F	11	
	2G	51	
	2H	75	
	2I	48	
Well Being (WB)	16C	73	
73	16D	73	