

Case Report

Habilitation of Child with Spinal Muscular Atrophy Type 2: A Case Report

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ABSTRACT

Spinal Muscular atrophy (SMA) is a severe neuromuscular disorder which is a hereditary genetic disease. It is an incurable disease due to a defect in the survival motor neuron 1 (*SMN1*) gene. This report shows the benefit of rehabilitation intervention for the habilitation of a child with SMA Type 2. A 5-year-8-month-old female diagnosed with SMA by genetic testing complained of an inability to stand independently. Her rehabilitation problems were muscle power function, mobility of joint function, respiration function, exercise tolerance function, mobility, self-care, school education, and economic life. We provided rehabilitation programs with a regular follow-up for six months, including flexibility exercises, breathing exercises, cardiopulmonary endurance exercises, standing support exercises, wheeling exercises, and activities of daily living (ADL) exercise. We coordinated with a psychologist and social workers to assess the psychology and socioeconomic condition of the patient and caregiver. The muscle power function, which was evaluated by functional motor ability assessment using the Revised Hammersmith Scale for SMA (RHS), didn't show any deterioration, and she had improvement in the ADL capability. We resolved the mobility of joint function problem in this patient for the joint stiffness in both knees, and Cobb's angle of scoliosis was decreased. Respiration function assessed by peak cough flow and chest expansion showed an increase, and exercise tolerance function was also increased. The patient received some donations from donators to overcome economic issues assisted by a social worker for the activity chair, standing frame, and modified wheelchair. She could mobilize well using her own modified wheelchair, played with friends, and attended school. A follow-up by the psychologist showed good psychological conditions of the patient and family. The quality of life assessment of patients by PedsQL Neuromuscular Modul showed improved results, and the caregiver burden assessment by The Zarit Burden Interview had decreased. Rehabilitation intervention for the habilitation of a child with SMA Type 2 can maintain, restore, and promote the patient's functional ability to provide a better quality of life and control the caregiver burden.

Keywords: *Quality adjusted life year, Rehabilitation, Spinal muscular atrophy.*

Introduction

Spinal muscular atrophy (SMA) is a severe neuromuscular disorder due to a defect in the survival motor neuron 1 (*SMN1*) gene.¹ The incidence of SMA is approximately 1 in 11,000 live births, and the prevalence is 1-2 persons per 100,000 worldwide.^{2,3} There is currently no official national report on the incidence and prevalence of SMA in Indonesia.³ The SMA community in Indonesia reported that 65 persons had been diagnosed with SMA in Indonesia. This disease is an incurable genetic hereditary and an autosomal recessive disease (the disease can manifest in both boys and girls). When both parents are carriers, each pregnancy has a 25% chance of producing a child with the disease.⁴ The gold standard of SMA genetic testing is a quantitative analysis of *SMN1* and *SMN2*. The absence of both complete *SMN1* copies will diagnose SMA, whereas *SMN2* copies are essential for prognosis and therapeutic approaches.¹

Spinal muscular atrophy includes a wide range of phenotypes classified into clinical groups based on the age of onset and maximum motor function achieved from SMA Type 1 (mildest) to SMA Type 4 (most severe).^{1,4,5} The SMA Type 2 is also called intermediate SMA, with an onset in 7-18

months of age, and the highest function achieved is sitting but not standing.⁵ The main objectives of the rehabilitation program for SMA type 2 are to prevent complications such as contractures and scoliosis and maintain and restore or promote function and mobility through habilitation.¹ The Committee of the Rights of Person with Disabilities (CRPD) define habilitation as "a process aimed at helping disabled people attain, keep or improve skills and functioning for daily living; its services include occupational, and speech-language therapy, various treatments related to pain management, and audiology and other services that are offered in both hospital and outpatients location."⁶

Contractures are common in patients with SMA due to decreased range of motion, prolonged static positioning, and agonist-antagonist muscle imbalance. Contractures inhibit function in patients with SMA both functionally and symptomatically. Inspection of the spine should be conducted as part of the routine clinical examination. Due to poor trunk and thoracic muscular support, children with SMA have an increased incidence of thoracic insufficiency due to scoliosis and distortion of the rib cage.¹

Table 1. Clinical Classification for Spinal Muscular Atrophy. Taken From Kolb et al.(2017)

	Age of onset	Highest function achieved
Type 1	0-6 months	Never sit
Type 2	7-18 months	Sit never stand
Type 3	>12 months	Stand and walk during adulthood
Type 4	2-3 decade	Walk unaided

Breathing problems were common for SMA types 1 and 2, but they are rare for types 3 and 4. It is recommended that everyone with SMA has both the annual influenza vaccination and the pneumococcal vaccine. All other routine vaccinations should be given as a recommended standard of care unless the health professional team advises otherwise.⁷ Pulmonary diseases are the leading cause of morbidity and mortality among patients with SMA types I and II.^{4,8}

The focus of the clinical assessment should be a physical examination supported by a clinical assessment of cough function. There is no clear consensus on the value of peak cough flow measurement or when should a sleep study be performed in the management of sitters.^{9,10} For sitters and standers, a consensus recommends that all patients able to perform spirometry should do so during each visit.⁹

A sleep study should always be

performed in symptomatic patients or when there is minimal suspicion of nocturnal hypoventilation.⁹ Sleep Disordered Breathing (SDB) is a common but under-diagnosed complication causing sleep disturbance and daytime symptoms in children with spinal muscular atrophy. A study in Europe showed the prevalence of SDB in SMA type 2 and 3 was 16.4%.¹¹ According to a statement by the American Thoracic Society, SDB in children is characterized by prolonged partial upper airway obstruction and/or intermittent complete obstruction (obstructive apneas) that disrupts normal ventilation during sleep and typical sleep pattern. In children with NMD, abnormal ventilation during sleep is primarily the consequence of the disease-related weakness of inspiratory muscles.¹²

The main topics of nutritional management in SMA include swallowing dysfunction and dysphagia, weight control, and gastrointestinal dysfunction. Over the last few years, there has been increasing evidence of possible metabolic abnormalities in SMA patients, such as altered fatty acid metabolism, impaired glucose tolerance, and muscle mitochondria defects.^{1,13-15} These findings suggest that SMN also affects particular enzyme production in the metabolism. Children with SMA may also suffer from various gastrointestinal problems, such as gastroesophageal reflux, constipation, abdominal distension, and retarded gastric emptying. It may result from abnormal gastrointestinal tract motility, reduced intake of foods rich in fibers and water, and hypotonicity of muscles in the abdominal wall.⁴ In all types of SMA, regular assessments of growth are essential. A nutritionist expert should be involved in providing an appropriate diet, monitoring weight and fluid, macronutrient, and micronutrient intake, especially calcium and vitamin D intake for bone health. Spinal Muscular Atrophy specific growth charts are not yet available. Secondarily to altered body composition in SMA, experts are divided on the use of standardized growth charts alone to monitor appropriate growth, but they may be helpful in monitoring trends.¹ For optimal care, nutrition evaluations are recommended after

diagnosis and periodically every 3–6 months for younger children and annual assessment afterwards. Chewing difficulties and fatigue with eating are frequent in sitters. Safe swallowing and the risk of aspiration are also a concern. History of choking or coughing episodes with feeds should be investigated and monitored with swallow studies. Feeding evaluations are also recommended for possible feeding modifications/occupational therapy to swallow safely and eat effectively. Longitudinal measures of weight and length in conjunction with body composition measures are recommended to promote appropriate growth.¹ Evaluation of feeding and swallowing includes: feeding assessment, mealtime observation, oral structures examination with positioning, and head control on feeding.⁸

Cardiac involvement is much less frequent in types 2 and 3 SMA. Recent studies performed in types 2 and 3 SMA suggested no need for regular cardiac surveillance in type 2 and 3 patients. It is improbable that these patients will develop noticeable clinical, ECG, or echocardiographic features of cardiomyopathy unless symptoms suggest a heart problem develops.^{7,9} All SMA type 1 and 2 patients should have wheelchairs with custom postural support and seating systems. Assessments for power wheelchair mobility can begin before two years of age. Lightweight manual wheelchairs or power assist wheels are ideal for promoting self-propulsion in more vital SMA type 2 patients to ensure functional independence. Exercise programs and activities that encourage muscle activation should be encouraged since they can affect maintaining and improving function, strength, range of motion, endurance, balance, activities of daily living, participation in school, social activities, and occupation.¹

Patients with SMA mostly have a cardiopulmonary endurance problem due to weakness and immobilization. There is no one-size-fits-all exercise approach to promote endurance and strength in SMA patients, and there is no recipe to follow. Aerobic exercise can be given, and it has been proven to encourage the motor function

and quality of life of SMA patients. According to previous studies, the optimal duration of individual bouts of aerobic exercise is ≥ 30 minutes, with a minimum frequency of 2 or 3 times per week and an optimal treatment frequency of 3–5 times per week.¹ The current aerobic exercise protocol for the SMA type II patients does not lead to severe fatigue or pain during and/or after exercise.¹⁶

Since the disease is incurable and may progress, the patient's quality of life and the caregiver's burden should be assessed. The PedsQL Neuromuscular Module demonstrates feasibility, reliability, and validity in the SMA population. It is completed both by children and parents to evaluate the child's quality of life. The instrument for parents contains the assessment of medical complications from neuromuscular disease, communication problems, and financial problems, while the tool for children only contains the functional disturbance that may present such as the difficulty of breathing, feeding, communication, mobilization and pain sensation.^{17,18}

Material and Methods

A 5 year 8 month old female presented to the rehabilitation clinic with a chief complaint of being unable to stand independently. She was diagnosed with global developmental delay at 1-year-old, with suspicion of SMA at 2-year-old, and confirmed SMA at 5 year 7 month old. She is the first daughter of two, and her little sister is three years old. Her father is an online driver, and her mother is a housewife. Both of them are high school graduates. There was no history of the same condition or SMA in the family. Her parents took her to the hospital for the first time when she was one year old because she couldn't sit independently. She was diagnosed with global developmental delay by a pediatrician, referred to a rehabilitation clinic, and received a rehabilitation program for one year. She had improved after one year of therapy. She could sit independently from the prone position and sit without support from 2 years old until now. She moved by scooting for 1-2 meters, and she felt tired afterwards. When she was

diagnosed with suspicion of SMA, her parents had stopped the medical treatment due to economic problems. They continued seeking medical treatment when she was five years old because her condition didn't improve, and she was eventually diagnosed with confirmed SMA. The pediatrician referred her to a rehabilitation clinic at the previous hospital and then referred her to the rehabilitation clinic in our hospital. Her parents were aware and had accepted her condition. She received free health insurance from the government for medical treatment. The parents wished that the patient could be independent.

She was cooperative during the examination. She could sit independently only from the prone position and sit without support for more than 1 hour. She moved by scooting for 1-2 meters, and she felt tired afterwards. She could read, write, and follow lessons at the kindergarten without any difficulty, except for physical education lessons.

The physical examination showed a regular respiratory rate and oxygen saturation. There was no bell-shaped deformity of the chest. The peak cough flow was 110 L/min with chest expansion of 1.5 cm on the upper thoracic, 1.5 cm on the middle thoracic, and 1.5 cm on the lower thoracic. The trunk alignment was a slight scoliosis c-curve that was worsened by fatigue. Adam's forward bending test on the sitting position was positive with the hump on the right side. The range of motions (ROM) was limited on both knees extension ($-5^{\circ}/-10^{\circ}$) with firm end feel (normal range $0^{\circ}/0^{\circ}$), and there was ligament laxity with elbow extension $25^{\circ}/50^{\circ}$ (normal range $0^{\circ}/0^{\circ}$). The manual muscle testing (MMT, score 0-5) of the extremities were all 4/4 except for the muscles of the shoulder 3/3, hip 2/2, and knee extensor muscles 2/2.

The functional motor ability using the Revised Hammersmith Scale for SMA was 30 (normal score of 69). The swallowing assessment was safe for all consistency. The ADL was assessed using WeeFIM and showed independence in self-care except for bathing, dressing, and toileting. The quality of life was evaluated using PedsQL Neuromuscular Module, with a score of 44 for the patient's report and 59 for the parent's

report (score range 0-100). There is no specific interpretation of the score; a higher score shows a better quality of life. The caregiver burden was assessed on her mother using The Zarit Burden Interview, which revealed little or no burden.

The Genetic testing showed *SMN1* deletion that suggested SMA. Radiological Examination for thoracolumbar AP-Lateral showed scoliosis thoracolumbar, with Cobb's angle 17°. The goal of the rehabilitation program for this patient was to promote the functional capacity to perform ADL more independently and prevent further complications such as scoliosis,

joint stiffness, and recurrent upper respiratory tract infection. The spirometry and capnograph test couldn't be performed due to the lack of facilitation at our hospital, so we conducted the sleep study for five days at home by asking the parent to check the pulse oximetry every hour during the sleep time. The lowest oxygen saturation during the test was 96%, with mostly 98%, which showed no sleep hypoventilation. Breathing exercise was provided with glossopharyngeal breathing and active chest expansion. She already had influenza vaccination once a year and pneumococcal vaccine.



Figure 1. First Thoracolumbar Radiologic Examination

The spirometry and capnograph test couldn't be performed due to the lack of facilitation at our hospital, so we conducted the sleep study for five days at home by asking the parent to check the pulse oximetry every hour during the sleep time. The lowest oxygen saturation during the test was 96%, with mostly 98%, which showed no sleep hypoventilation. Breathing exercise was provided with glossopharyngeal

breathing and active chest expansion. She already had influenza vaccination once a year and pneumococcal vaccine. Since she felt tired easily, we gave her aerobic exercise to increase her cardiopulmonary endurance. She received arm ergo cycle exercise twice a week at the hospital with duration and intensity tolerated until moderate to fatigue (Borg scale 13-16).



Figure 2. Aerobic Exercise Using Arm Ergocycle

The patient received flexibility exercise by active assistive ROM for lower extremities with stretching knee flexors twice a week by a physiotherapist at the hospital and the caregiver at home twice a day. The vigorous assistive ROM exercise was administered 30 times each session, while the stretching exercise was administered ten times each session, and she held her position for 30 seconds. After the ROM was complete, we gave a standing facilitation exercise using a standing frame at home once a day for 30 minutes of duration.

Activities of the daily living exercise were given by an occupational therapist at the hospital twice a week for dressing, toileting, and bathing independently with modifications, such as choosing the shirt type as her clothing for easier dressing and

bathroom modification by providing a sitting toilet and shower. We also gave education for performing all ADLs in a sitting position using an activity chair to promote good posture.

We coordinated with the social worker to find a donator for the standing frame, activity chair, and modified wheelchair with a light frame, body strap, and portable tray. She received wheeling exercise using her modified wheelchair at the hospital twice a week. Because she would enter the elementary school in a couple of months, we also coordinated with the social worker to find a suitable school for her. The psychologist suggested that she enter an ordinary elementary school. We also referred her to a pediatrician for further assessment of her nutritional status and needs.

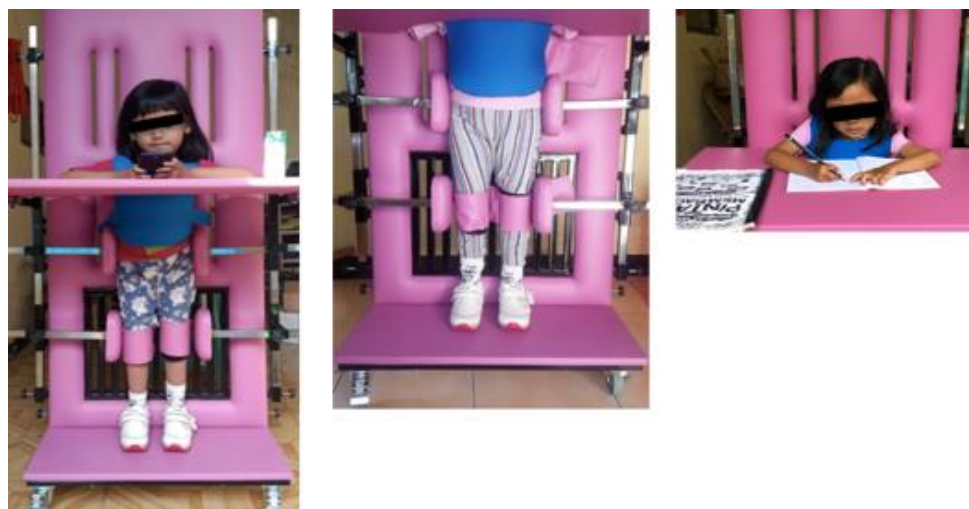


Figure 3. Standing Facilitation Exercise at Home

Result

After five-month exercises, there was no deterioration of RHS from the previous examination. The peak cough flow was increased to 140, and chest expansion became 3/3/3 cm. The ROM of all joints became full. The Cobb's angle on the thoracolumbar radiologic follow-up examination was decreased to 12°.

The WeeFIM score was increased by 8 points in upper dressing, toileting, and bathing. There was also improvement in aerobic exercise duration using the arm ergocycle, from 10 minutes to 30 minutes, before the patient felt tired. The patient already received donations for the standing frame, modified activity chair, and modified

wheelchair. The patient could use standing support at home for 30 minutes without complaint.

The patient could wheel independently for 200 meters in 30 minutes in and around the house before feeling tired. The quality of life assessment using the PedsQL Neuromuscular Module was increased in the patient's report to 64, and the caregiver burden was still stated as little or no burden. The pediatrician had already assessed the patient's cardiac status and said no cardiac involvement. The nutrition status assessment from pediatric nutrition and metabolic evaluation stated that she had a good nutritional status and already described her dietary requirement.



Figure 4. Modified Activity Chair



Figure 5. Modified Wheelchair

Discussion

The patient had joint stiffness in both knees due to prolonged immobilization and weakness of knee extensor muscles. We provided flexibility exercises using manual stretching until the ROM became full, and then we offered supported standing exercises. The supported standing exercise facilitates lower-extremity stretching and promotes bone health, enables upright participation, and improves trunk posture in those with sufficient lower extremity range of motion.^{1,7} Optimal duration for standing is ≤ 60 minutes, minimum frequency 3–5 times per week, and optimal treatment frequency 5–7 times per week.⁷ The support standing is also facilitated with a portable tray, so the patient can do activities while exercising.

We gave conservative treatment for her scoliosis by setting an adaptive seating system on the activity chair. For SMA type 2 patients, scoliosis $<20^\circ$ should be monitored every six months until skeletal

maturity and yearly after skeletal maturity. Management with spinal orthoses is often advocated to support the hypotonic trunk and treat scoliosis $>20^\circ$, especially in a child with remaining significant growth.¹ For sitters or patients who use wheelchairs with scoliosis, a customized, adaptive seating support system with 3 points pressure system using external lateral trunk support (adjusted lateral trunk support pads) can improve sitting posture. Such supports can be applied based on a 3-point force system, an engineering concept in which two parallel forces are opposed by force acting in the opposite direction. The adaptive support system is more effective and favourable to prevent further progression than a spinal brace. The latter causes significant chest and/or abdominal discomfort leads to lack of compliance, and risks pressure injury.⁸ There is still no evidence-based literature regarding the recommended duration of seating for each day in SMA patients. Still,

pressure relief by regular positioning every 15-30 minutes for 15-120 seconds is recommended to prevent pressure injury.¹⁹

The spirometry and capnograph test couldn't be performed in our hospital due to the lack of facility, so we conducted the sleep study at home. Oximeters can evaluate sleep hypoventilation at home by overnight pulse oximetry with manual chart recording.^{5,10} An acute decrease in oxygen saturation to <95% while asleep suggests hypoventilation or mucus plugging.⁹ Hypoventilation is usually associated with long-term oxygen desaturation. A decreased oxygen saturation of <90% for >5 minutes with a nadir of ≤85% may also indicate hypercapnia and should urge the clinician for further diagnostic workup.²⁰ The lowest oxygen saturation on this patient was 96%, mostly 98%. We still had concerns about the respiration in this patient, although the sleep study showed no oxygen desaturation event (oxygen saturation <90%) since she had decreased chest wall compliance which could lead to inadequate lung expansion.⁸

The feeding assessment in this patient was safe for all kinds of textures. The weight and height growth charts were within the normal range. To ensure that the patient has adequate nutrition and prevents gastrointestinal problems, we referred her to pediatric nutrition and metabolic expert. Optimal nutritional management for patients with SMA includes longitudinal evaluation of weight and length, dietary analysis, and preventing constipation. There is still no consensus or research that determines the specific recommended type of food or precaution in SMA. Further research is needed to assess the use of elemental and semi-elemental formulas in SMA management, including the optimal intake of macronutrients and micronutrients for nutritional support and the ideal fat content and composition.¹⁵

We provided a modified light wheelchair to promote her mobilization and function because MMT for her upper extremity was capable of wheeling. But we still considered the patient's endurance for long-distance wheeling since she complained of being easily fatigued. We provided aerobic exercise to increase her cardiopulmonary endurance capacity. To

avoid dull feelings and increase their enjoyment, we give her exercise while watching her favourite film on her mobile phone. We also educated the patient to stay as active as possible, as long as she enjoys doing the activities. We didn't perform a 6-minute arm ergometer test (6-MAT) on this patient before and after the exercises, but she showed progression in duration tolerance each exercise. The 6-MAT is a submaximal exercise testing for assessing VO₂max prediction of individuals with lower limb disorders. Still, it is not commonly used since there is no standardization of the VO₂max value in SMA patients.²¹ Some previous studies showed that ergometer training for 12 weeks with moderate intensity effectively improved motor functions, exercise performance, and quality of life. Arm and leg ergo cycle can be used safely among children with SMA type 2, as in other kinds of SMA.^{16,17}

Because SMA is an incurable disease, the optimal quality of life is something that we should promote. The PedsQL neuromuscular module can be used to evaluate the parameters in SMA patients. PedsQL in the patient's report showed improvement. Better functional ability and quality of life in this patient also can decrease the caregiver burden.^{17,18}

Conclusion

Rehabilitation intervention for the habilitation of a child with SMA Type 2 can maintain, restore, and promote the patient's functional ability to provide a better patient quality of life and control the caregiver burden.

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