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Letter to Editor

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Spinal Muscular Atrophy Management in Bangladesh

Dear Editor,

Spinal muscular atrophy is an inherited progressive neuromuscular disorder characterized by muscle atrophy and weakness due to loss of spinal cord motor neuron. SMA is a rare disease and its incidence is 1in every 10000 to 11000 livebirths worldwide. Carrier of SMA is 1in 54 people. Clinical presentation of SMA is variable ranging from severe disability to mild impairments. According to age of onset and clinical presentation SMA is classified into 4 types. Type I SMA is most lethal, presented with severe motor delay and respiratory distress and most of patient die before their first birth day. Type II presented at the age of 6 months with severe disability and recurrent respiratory infection and patient never achieved independent standing, type III face moderate difficulties in walking with normal life span. Type IV is adult onset and has mild impairment.

SMA is an autosomal recessive genetic condition caused by the deletion of two pairs of SMN 1 genes, which are needed to produce SMN protein. The survival of motor neurons depends on the SMN protein, and the severity of the condition is determined by the supporting two pairs of SMN2 genes, which produce a little amount of SMN protein. Insufficient production of SMN protein can therefore lead to a progressive loss of motor neurons, which can cause skeletal deformities, difficulty in eating, and breathing, and muscle weakness even death. SMA is rare and non-curable disabling disease, before 2016 there was no specific treatment, patient was managed symptomatically. Today we have three option to improve their condition that is gene replacement therapy zolgensma and two gene directed therapy nussinersin, rizdiplam. All three molecule are effective, improve their muscle power, quality of life, but very costly. It is nearly impossible for parents to afford the cost. Without appropriate management most of patient die within short period of time, or may grow up with disability. This illness has a significant effect on society and the health industry. The patient's family suffers from the high expense of treatment and repeated hospital stays. Raising this disabled child is incredibly painful for the family.

In this situation we have to pay attention to some key aspects which help to improve facilities of SMA in Bangladesh. In Bangladesh, people are less aware of rare disease like SMA. The general state of SMA management can be improved with awareness. We have no appropriate health infrastructure for SMA to provide appropriate care and confirm the diagnosis. This can lead to delay in diagnosis appropriate management care. Bangladesh like other LMIC this burden particularly challenging. It is extremely challenging for parents to manage a SMA patient with expensive medication and other supporting care. For this reason, many are hesitant to get the treatment. As lack of knowledge and awareness support services for SMA patient and family are limited. This could include access to support groups, specialized centers and caregiver. As a resource poor country research and development in the field of rare disease like SMA may not receive as much as attention or funding compared to other prevalent disease. Advocacy effort may be necessary to raise awareness about SMA among policymakers and steak holders. This could help to prioritize the needs of development of support system to manage the SMA patient.

The perspective of SMA in Bangladesh likely involves challenges related to awareness, infrastructure, support system, research and advocacy. Addressing these challenges would require a multifaceted approach involving and collaboration between Government and healthcare professional advocacy group and international organization.

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