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# Collaborative program for the management of a patient with Mucopolysaccharidosis VI : A three-year follow-up

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## Introduction

Mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome) is a rare genetic lysosomal storage disease resulting from a deficiency in the lysosomal hydrolase N-acetylgalactosamine-4-sulphatase (4S). This enzyme deficiency leads to multisystemic abnormalities with numerous clinical manifestations including respiratory and cardiovascular complications, gastrointestinal symptoms, restricted joint movement, and loss of vision and hearing. MPS VI is a progressive disease which leads to severe functional disability and a shortened life span in most cases. In view of this, a collaborative program with Department of Paediatrics was launched since 2009. The program includes regular Enzyme Replacement Therapy (ERT) as prescribed by Pediatricians. Physiotherapists perform a comprehensive physical assessment and accordingly prescribe a home exercise program with care advice. Regular follow-ups were done to ensure patients complied with prescribed exercises.

## Objectives

To evaluate the effectiveness of a collaborative program on the physical and functional abilities of a patient with MPS VI over a 3 year course

## Methodology

The physical and functional abilities of a 20 years old male with advanced MPS VI were assessed by a physiotherapist from July 2009 to June 2012. The patient received weekly infusions of recombinant human arylsulfatase B and regular physiotherapy intervention throughout the study. Outcome measures included 6-minute walk test (6MWT) and 3-minute stair climb (3MSC)

## Result

Assessment analysis revealed a great improvement in both the 6MWT and 3MSC test with a 227% and a 52.8 % increase respectively. The exercise compliance of the patient was satisfactory and the patient reported that the home exercise program and advice was useful to him. He also stated that he was more willing to perform home

exercise and with a better understanding of his physical condition. The collaborative program for patients with MPS VI was found to be useful to them in terms of improving their physical and functional abilities. Moreover, the patient engaged in a more active and healthy lifestyle through a change of exercise habit and better understanding of his own physical condition.