INTRODUCTION: Type I mucopolysaccharidosis is a lysosomal disease, classified into severe and attenuated forms. Although for the severe form hematopoietic stem cell transplantation (HSCT) is recommended whenever the diagnosis is made before 36 months, in practice few patients perform HSCT. OBJECTIVES: To review and describe the steps involved in the indication, performance and follow-up of patients with severe MPS I undergoing HSCT in Brazil, and to evaluate their results. MATERIALS AND METHODS: A cross-sectional review study was carried out at the Medical Genetics Service of the Hospital de Clínicas of Porto Alegre, with the participation of the collaborating centers of the MPS Brazil Network, from March 2015 to January 2018. 9 patients were included. The variables evaluated were: transplantation center, age at diagnosis, mutations, type of donor, use of enzyme replacement therapy, age at transplantation, outcome after HSCT. The data were compared with those available in the international literature. RESULTS: the mean age at transplantation was 3.7 years; however, mean age was reduced to 1.8 years when we considered the 8 patients who were transplanted after 2010. Considering these 8 patients, the age at diagnosis was 9 months, and the time between diagnosis and the first transplant ranged from 5 to 18 months. Conclusion and discussion: Neutrophilic time was comparable to those reported in the literature, which may demonstrate that the technique of HSCT is in agreement with the overall experience. Although diagnoses were made early, there was a large gap between diagnosis and HSCT procedure. The delay between diagnosis and the first HSCT may have contributed to the high mortality rate (4/9 patients died due to procedure complications) and low efficiency (6/9 had an unfavorable treatment outcome). In Brazil, 301 patients were diagnosed with MPS I so far and, to the best of our knowledge, only 9 performed HSCT. These data reiterate the need to take measures that may contribute to improve the efficiency of HSCT in MPS I in Brazil, in order to obtain favorable results closer to those observed in North America and Europe (above 80-90% of survival and favorable outcome).