Immunisation issues in patient with mucopolysaccharidosis: A case report

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SUMMARY

Young patients, especially infants with Mucopolysaccharidosis (MPS) have increased risk of recurrent upper and lower respiratory tract infections. A complete schedule of immunisations is crucial to protect children from life-threatening infections. However, in most cases, they often miss scheduled vaccinations due to many factors. This case report describes issues in administering routine immunisations to infants with MPS. It is vital to recognise the indications and contraindications of vaccinations for patients with MPS although all vaccines need detailed study to investigate their safety and immunogenicity. Furthermore, regular educational programs are essential for both parents and health providers.

INTRODUCTION

The Global Vaccine Action Plan (GVAP) aims to extend the benefits of immunisation equitably to all people. This includes coverage for the high-risk groups of vaccine-preventable diseases, such as preterm infants, chronic diseases, and immunocompromised patients, among others. However, despite the increased publicity and provision of funding for immunisation programs as well as the continuous promotional efforts, vaccination delays or missed opportunities for immunisation still commonly occur in children with special condition such as rare diseases.[1]

Mucopolysaccharidosis (MPS) is a rare disorder encompassing a group of a metabolic disorders caused by defective lysosomal storage. Ear, nose, throat (ENT), and respiratory problems are the first symptoms to present in infants with MPS and are very commonly found in young patients.[2] The issue is complicated since this particular population of patients are often under-vaccinated for various reasons. The lack of awareness of vaccine-preventable diseases, and doubts or misunderstanding concerning the safety and efficacy of vaccinations among patients, parents, and healthcare providers contribute to the low coverage of immunisation in this group.[3]

This case reports about a 3-month-old infant who suffers from the rare genetic disease mucopolysaccharidosis, who had missed routine vaccinations in the immunisation schedule, which should be administered according to his age.

CASE REPORT

A 3-month-old male infant was referred to our outpatient department for immunisation. The patient had been diagnosed with MPS type two (MPS-T2). Medical history reported he was born to a 32-year-old mother at 34 weeks of gestation by spontaneous vaginal delivery. His Apgar scores at one and five minutes were five and seven, respectively. Physical examination revealed a child of a short stature, enlarged skull, low nasal bridge, thick coarse facies, short hands and small feet. Before being discharged, the first dose of Hepatitis-B vaccine was administered. There were no adverse events reported afterwards. The parents were recommended to seek routine medical check-ups for assessing the growth and development of the child and continuing the immunisation according to the provided schedule.

At two and a half months, the parents took the child to the local primary care centre for administering the Bacillus Calmette-Guerin (BCG) and the first dose of the Inactivated Polio Vaccine (IPV). However, the health providers refused to administer the immunisation on schedule. They were afraid that the vaccine could have a side effect, or it might worsen any underlying conditions. The primary care facility referred this patient to our centre and to obtain a recommendation letter for the immunisation. On that visit, the condition of the patient revealed no contraindications for the immunisations. Accordingly, the BCG, and the first dose of the IPV vaccine were subsequently administered. This patient was observed for one hour after immunisation, his condition was found to be stable and no adverse event following immunisation was reported. With the recommendation letter that we provided; the patient could receive further immunisations at the primary care centre near their home.

DISCUSSION

There are no particular recommendations regarding the standard vaccinations for children with rare diseases. One study was conducted in Italy that evaluated the immunisation coverage in children with three different rare genetic diseases. It showed that compared to the healthy controls, the coverage of scheduled recommended vaccines in children with genetic syndromes was significantly lower.[1]

The diagnosis of MPS involves a group of inherited metabolic diseases, which are caused by the lack or malfunction of certain enzymes. These patients with underlying metabolic

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disorders are at a potentially high-risk group for vaccine-preventable diseases.[4] The benefits of vaccine-associated immunity far outweigh the risks related to vaccine administration. Routine vaccinations can protect them from the more severe symptoms due to natural infection. Therefore, the American Academy of Pediatrics Committee on Infectious Diseases recommends that children with metabolic disorders receive the same scheduled immunisations as per healthy children. Except there is a contraindication to specific components of the vaccines. In-depth data related to the risks and immunogenicity of each vaccine for this unique population are still necessary to define the exceptions, and to determine whether specific dosages and schedule are required for those patients.[5]

On the other side, many factors contribute to the low coverage of vaccines for patients with MPS and other genetic disorders. The first is a lack of knowledge about the importance of the scheduled vaccines. People who live in a developing country with population with lower education levels will have minimal information about the importance of immunisations. Some also refuse the vaccine because of their religious and cultural beliefs. Many parents and health providers still worry about' the safety and effectiveness of some vaccines, whether they will be harmful rather than provide any benefit. There are concerns about the adverse effects of some vaccines, especially in children with special conditions. ' Fear of vaccines among parents can worsen the existing condition leading to vaccine refusal.[1] Similar issues were also faced by other patients with genetic diseases such as Alagille syndrome, Cornelia de Lange syndrome, and others. For these cases, primary care providers also need a recommendation note from a specialist before they administer the scheduled vaccines.

Accordingly, a health provider needs to build trust with parents by increasing their knowledge and giving them confidence to make informed decisions. In this partnership, both sides are responsible to increase compliance and the coverage of the scheduled immunisation to all population, especially in children with a vulnerable condition.

A thorough, meticulous study of the safety and immunogenicity of vaccines related to all metabolic diseases would be a challenging task. A vaccine database can initiate this important registry for children with metabolic diseases and/or other genetic diseases. It will provide a valuable source of data to inform health providers and parents when they are making decisions about the relative safety and benefits of specific vaccines for their vulnerable children with metabolic disorders.[5]

CONCLUSION

This is a case report of an infant who suffers MPS, a rare genetic disease. Due to a lack of knowledge among health care providers, the patient had missed some vaccinations in the immunisation schedule, which should be administered according to his age. This case emphasizes the importance of recognising the indications and contraindications of immunisation for the patients with MPS. Further careful study of the safety and immunogenicity of any vaccine in all metabolic diseases is needed, and a vaccine registry would certainly be a valuable source of current information. Educational programs are equally essential for both parents and health providers, and scheduled vaccinations should be promoted as part of the consultation throughout routine appointments with the specialists. It is vitally important to increase the awareness among caregivers and health providers of the value of timely vaccines related to these special conditions.

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