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#13586 Summary

SUMMARY REVIEW EDITING

Submission

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

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Title and Abstract

Title THALASSEMIA β MAJOR WITH EXTRAHEPATIC CHOLESTASIS AND CHOLELITHIASIS IN GIRL 11 YEARS 9 MONTHS
Abstract

Kolestasis dapat disebabkan oleh gangguan fungsional hepatosit dalam sekresi empedu dan/atau karena obstruksi pada setiap tingkat jalur ekskresi empedu. Pada pasien thalassemia terjadi hemolisis kronis yang berakibat bilirubin tak terkonjugasi akan mengkristal dan akhirnya membentuk batu. Selanjutnya dapat terjadi penumpukan bilirubin disaluran empedu sehingga kemudian menyumbat dan terjadi gangguan pengeluaran bilirubin direk sehingga terjadi kolestasis. Kabaruan penelitian ini adalah meneliti kejadian kasus Thalassemia Mayor Dengan Kolestasis Ekstrehpatis Dan Kolelitiasis Pada Anak Perempuan 11 Tahun 9 Bulan. Tujuan penelitian ini untuk menggambarkan kejadian kolestasis dan kolelitiasis yang terjadi akibat proses hemolisis pada thalassemia β mayor dengan keluhan utama nyeri perut dan ikterus pada seluruh tubuh. Pasien didiagnosis berdasarkan temuan laboratorium yakni peningkatan enzim transaminase, bilirubin direk dan pemeriksaan multislice computerized tomography abdomen dengan kontras. Tatalaksana pasien ini dilakukan endoscopy retrograde cholangiopancreatography dan asam ursodeoksilat disertai terapi thalassemia. Kesimpulan penelitian ini memiliki prognosis yang baik karena respon yang baik. Meskipun kolestasis jarang terjadi pada pasien thalassemia pada anak-anak, dokter anak harus menyadari hal ini dan pengobatan segera harus ditangani.

Kata kunci: Thalassemia; Kolestasis ; Kolelitiasis.

Abstract

Cholestasis can be caused by functional impairment of hepatocytes in bile secretion and/or by obstruction at any level of the bile excretory pathway. In patients with thalassemia, chronic hemolysis occurs, which results in unconjugated bilirubin crystallizing and eventually forming stones. Furthermore, there can be a buildup of bilirubin in the bile duct so that it clogs and interferes with the release of direct bilirubin, resulting in cholestasis. The novelty of this study is to examine the incidence of cases of Thalassemia Major with Cholestasis Extrhepatis and Cholelithiasis in 11 Years 9 Months Girls. The purpose of this study is to describe the incidence of cholestasis and cholelithiasis that occur due to the hemolysis process in thalassemia β major with the main complaints of abdominal pain and jaundice throughout the body. The patient was diagnosed based on laboratory findings, namely elevated transaminase enzymes, direct bilirubin, and multislice computerized tomography of the abdomen with contrast examination. The management of this patient was endoscopic retrograde cholangiopancreatography and ursodeoxylic acid, accompanied by thalassemia therapy. Conclusion of this research has a good prognosis because of the good response. Although cholestasis is rare in thalassemia in children, pediatricians should be aware of this and seek immediate treatment.



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References

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