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CEFAZOLIN INDUCED STEVENS - JOHNSON SYNDROME IN PAEDIATRIC PATIENT WITH MULTIPLE VENTRICULAR SEPTAL DEFECTS & SEVERE PULMONARY HYPERTENSION

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ABSTRACT

To report a case involving Steven Johnson syndrome as ADR for cefazolin while surgical prophylaxis in a paediatric patient. There are several drug which induce SJS as a severe ADR, which is characterized by immune-complex–mediated hypersensitivity that typically involves the skin and the mucous membranes, toxic epidermal necrolysis, with body surface area (BSA) detachment . Stevens–Johnson syndrome has long been considered to resemble erythema multiform with mucosal involvement, but is now thought to form a single disease entity with toxic epidermal necrolysis. Although Stevens–Johnson syndrome is less severe, etiology, genetic susceptibility and patho-mechanism are the same for Stevens–Johnson syndrome/toxic epidermal necrolysis. The condition is mainly caused by drugs, but also by infections and probably other risk factors not yet identified. Identification of the cause is important for the individual patient and in cases of drug-induced disease withdrawal of the inducing drug(s) has an impact on the patient's prognosis. A majorly prescribed drug to prevent bacterial infections and surgical prophylaxis is cefazolin i.e. II cephalosporins, induces SJS in case, if prescribed for surgical prophylaxis.

INTRODUCTION

Stevens-Johnson syndrome is both a physically and psychologically devastating disease[1]. This paper primarily deals with the physiological complications of the disease process, but the psychological trauma often associated with such an initially disfiguring disease leaves wounds that are not visible[2]. Constant support of both the patient and the nursing staff is necessary to relieve some of the anxiety associated with this syndrome.[3] Education and reassurance should be as much a part of the treatment process as drug therapy.

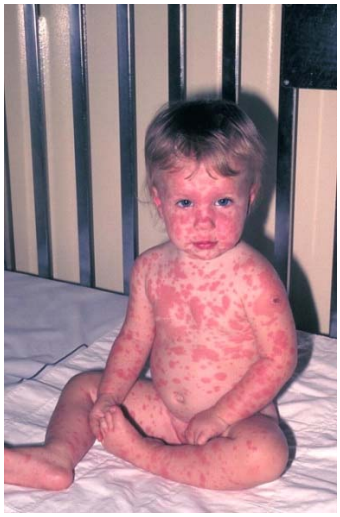
Although minimal drug therapy is required in the treatment of Stevens-Johnson syndrome, early aggressive management is necessary.[4] Clinical presentation and manifestations of SJS include the skin, eyes, gastrointestinal tract, and pulmonary system[5]. Infectious complications are the leading cause of mortality. Early intervention is important to prevent progression of SJS[6]. The case described is consistent with the features of this syndrome. The patient presented with fever, arthralgias and malaise[7]. Skin lesions included a diffuse violet macular rash with erythema and multiple bullous lesions on his neck and abdomen. [8]The skin biopsy was consistent with SJS. Multiple mucocutaneous ulcers were found in her mouth, but no evidence of lower gastrointestinal tract involvement was documented[9]. Treatment should include management of pain and fluid losses as well as supportive care of the respiratory and ocular complications. It is essential that nutrition be maintained and that treatment of infections is appropriate to the identified cultures. [10]Antacids, H_2 receptor antagonists, or both have proven beneficial in the prevention or reduction of gastrointestinal ulcers. Most important, however, is psychological support of the patient.

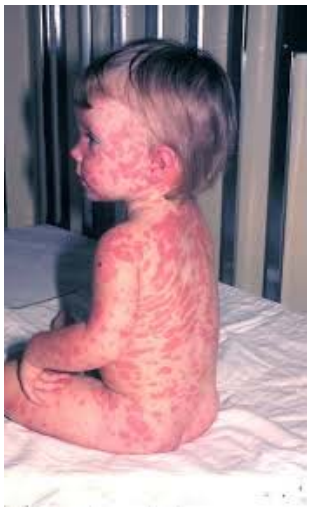
CASE REPORT

In a case of 2yrs paediatric male patient who underwent pre-operative cardiac catherization to see reversibility of PAH (pulmonary arterial hypertension) and inj. Cefazolin 500mg i.v. (bd) was given on 13/07/2013 for surgical prophylaxis as multiple VSD closure for VSD (ventricular septal defect) was planned on 14/07/2013.the concurrent drugs in prescription included – tab. Furosemide 40mg p.o , tab. Spironolactone 25mg p.o, nasoclear saline nasal drops 2 drops t.i.d., syp. Chlorpheniramine maleate 2.5ml b.d and inj. Heparin 100mg stat..

He developed multiple polymorphic rash all over the body involving palms and soles, also oral mucosa with peeling of skin. In the view of suspected SJS, dermatology consultation sought who stated on pulse methyl prednisolone followed which he recovered well. After full recovery multiple VSD closure was done on 10/08/2013.

The perimembrane VSD was repaired with another pericardial patch using 5/0 prolene, continues suture.ASD closed directly.the tv was tested and found competent. A closure was done. heart weaned of CBP with stale haemodynamics.in intra-operative TEE showed no significant residual VSDs.decanalation done,heamostasis chesked,clest closure done.





Post-operative period was uneventful. He was ventilated over night with good haemodynamics.at the time of discharge he was comfortable, on regular feeds. The wound healed well, his chest x-ray and 2D-echo showed no major issues.

DISCHARGE ADVICE

DOSAGE FORM	DRUG	ROUTE	DOSE	FREQUENCY
Tab	Furosemide	p.o	40mg / 5ml of water	t.i.d
Tab	Spironolactone	p.o	½ tab / 5ml water	b.d
Tab	Sildenafil	p.o	4mg / 6ml of water	q.i.d
Tab	Enlapril	p.o	1.25mg / 5ml of water	t.i.d
Nasal drops	Nasoclear	i.n	2 drops each nostril	6^{th} hrly
Sachet	Laxopeg		2 sachets	s.o.s

CONCLUSION

In order to avoid morbidity and mortality associated with SJS, it is at most significance to be vigilant while giving drugs known to cause SJS, early diagnosis, identification of the culprit drug, its prompt withdrawal and specialised supportive care. Since, cefazolin has been reported to cause SJS.its use for cardiac catherization prophylaxis needs to be reconsidered in the view of safer alternatives available.

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