



Olgularla İntradiyalitik Komplikasyonlar

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Sare

5 ay kız hasta

Konjenital nefrotik sendrom

Öykü

- 38 GH, term, 3000 gr
- Kuzen evliliđi
- 2 aylık ödem
- 4 aylık SDBY ve PD
- Mantar peritoniti nedeni ile HD

Başvuru

- Boy: 61 cm (-1.3 SD)
- Tartı: 4.75 kg (-3.3 SD)
- BÇ: 38 cm (-3.1 SD)
- KB: 95/62 mmHg

- Anürik
- NG ile besleniyor
- Sık enfeksiyon
- Hipotiroidi

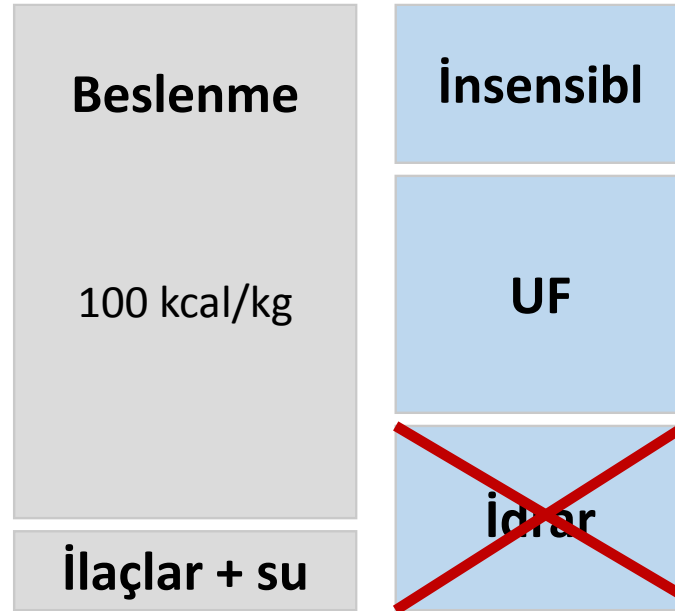


Sare - Diyaliz Reçetesi

Diyaliz Türü	HD
Damar giriş yolu	6.5 f HD kateteri
UF hedefi	?
Diyalizör	0.2 m ²
Set	Fx Paed/Baby
Süre/Seans	4 sa/4 gün
Na	140 mEq/lt

Sare, 4 kg

%5 VA/seans veya 10 ml/kg/saat



Sare – İnradialitik sorunlar

Hipotansiyon

Yetersiz UF

Kardiyovasküler risk

Semtomatik Hipoglisemi

Terleme, taşikardi

Konvulsiyon

Diyalizde hipotansiyon sebepleri

Sepsis

Serebrovasküler olay

Aritmi

Tromboemboli

Hemoraji

Hızlı UF

Aşırı IDWG

Diyaliz öncesi
Antihipertansif kullanımı

Beslenme

Anemi

Diyalizde hipotansiyonun sonuçları

Kasılmalar, ağrılar

Kronik hipervolemi

Yetersiz diyaliz

RRF azalması

Geçici iskemik atak

Serebral perfüzyon ↓

«Şaşkın miyokard»

Aritmi

Miyokard iskemisi

Diyalizde hipotansiyon tedavisi

Beslenme \emptyset

Antihipertansif \emptyset

Na profili

RBV kullanımı

Diyaliz programı

Süre

Seans

Midodrin

Hipoglisemi

- Inborn error of metabolism
 - Glycogen storage diseases
 - Fatty acid oxidation disorders
 - Ketogenesis disorders
- Endocrine disorders
 - Hyperinsulinemia
 - Adrenal insufficiency
 - Steroidogenesis disorders
 - Adrenal damage
 - Peroxisomal disorders
 - Abnormal adrenal development
 - Adrenal unresponsiveness to ACTH
 - Growth hormone deficiency
- Chronic kidney disease/hemodialysis associated
 - Malnutrition (depleted glycogen stores)
 - Low alanine/glutamine blood concentration
 - Cessation of enteral feeding
 - Carnitine deficiency
 - Catheter-related infections
- Others
 - Sepsis, shock
 - Liver dysfunction
 - Medication (beta-blocker, salicylates, steroid withdrawal)

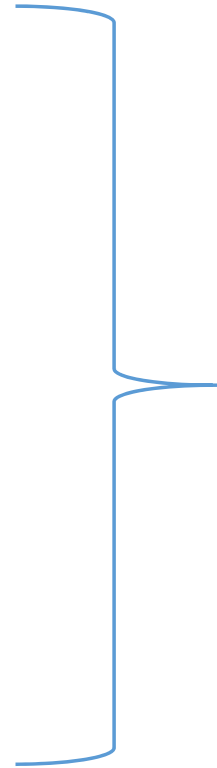
Sare - Diyaliz Reçetesi

Diyaliz içi enteral
beslenme kesildi

Intradiyalitik parenteral
beslenme
GPH 4 mg/kg/dk

Glukozlu diyaliz solüsyonu

Diyalizat Na ↑



İnatçı
Hipoglisemi

Tanı ?

1
3
2

CLINICAL QUIZ

4

5

6

Persistent hypoglycemic attacks during hemodialysis sessions in an infant with congenital nephrotic syndrome: Answers

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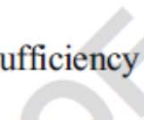
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Keywords Congenital nephrotic syndrome · Hypoglycemia · Hemodialysis · Adrenal insufficiency · *SGPL1* mutation





Teşekkürler!

Sphingosine-1-phosphate lyase mutations cause primary adrenal insufficiency and steroid-resistant nephrotic syndrome

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Mutations in sphingosine-1-phosphate lyase cause nephrosis with ichthyosis and adrenal insufficiency

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