## In the Name of God

Follow-Up

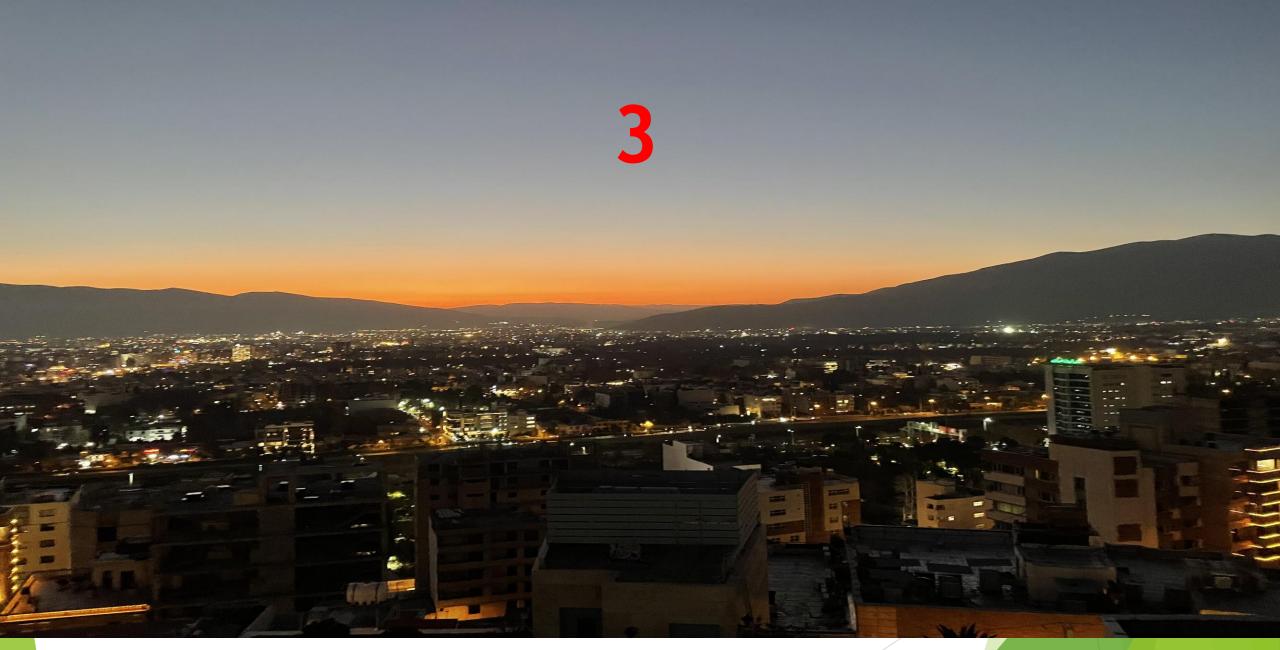
M. Haghighat

Professor of ped. Gastroenterology and Hepatology

SUMS, Shiraz, IRAN, 1403



طلوع و غروب خورشید در شیراز در یک روز



شيراز



شيراز





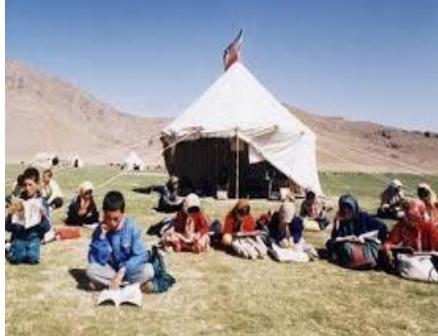






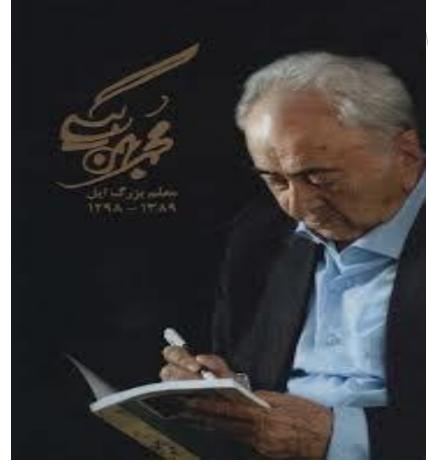






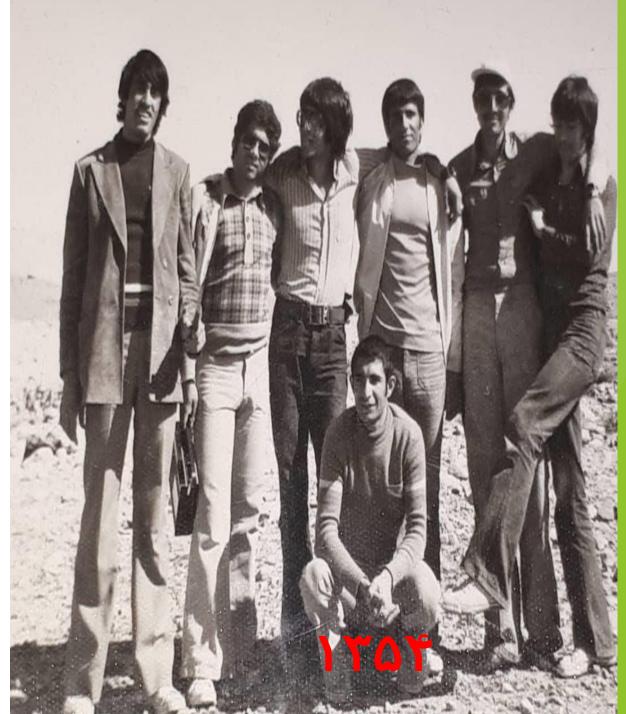


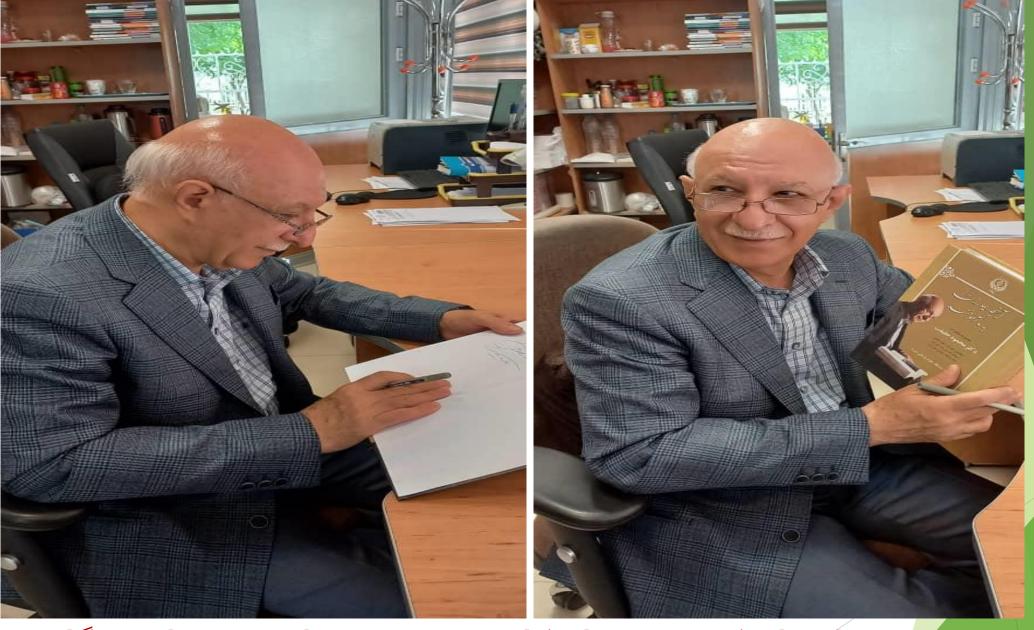




استاد محمد بهمن بیگی , بنیانگذار آموزش عشایر ایران







خدایاً چنان کن سرانجام کار تو خشنود باشی و ما رستگار







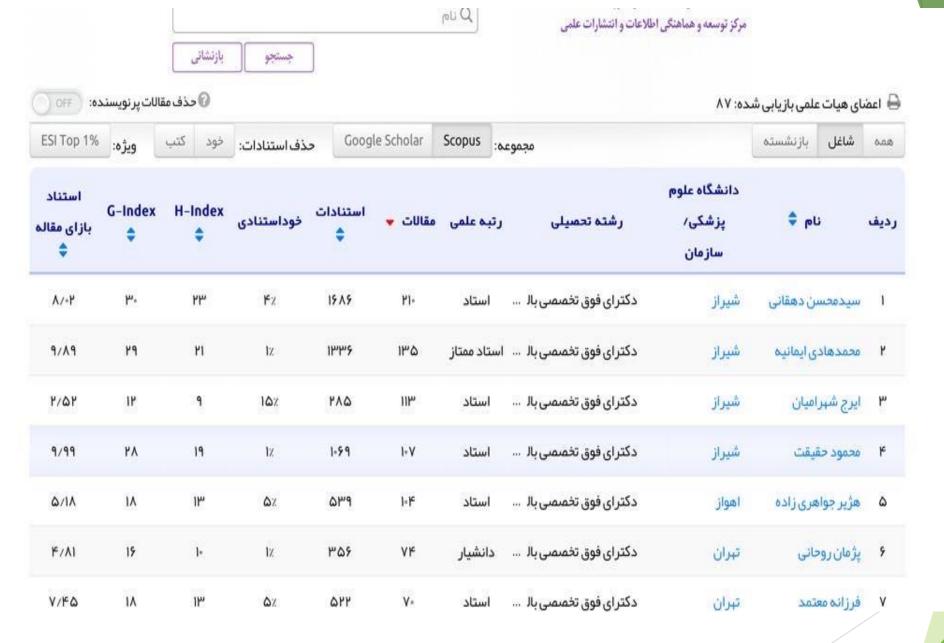
سامانه علم بجى اعضاى هيات علمى

وزارت بهداشت درمان و آموزش پزشکی معاونت تحقیقات و فناوری مرکز توسعه و هماهنگی اطلاعات و انتشارات علمی

رتبه علمي	هیات علمی	*
دکترای فوق تخصصی بال ×	گوارش کودکان ×	×
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سامانه علم سنجی بر اساس H-Index



سامانه علم سنجي براساس مقالات











دکتر محمدحسین انباردار



تجلیل از پرستاران بخش های گوارش و اندوسکوپی

# نگاهی به زندگی مادر ترزای ایران

مشاغلی با درآمدهای بسیار بالا از سو ی مراکز پزشکی تراز اول اروپا و آمریکا به او پیشنهاد می شد اما او هیچگاه تحت تاثیر این پیشنهاد قرار نگرفت و ...

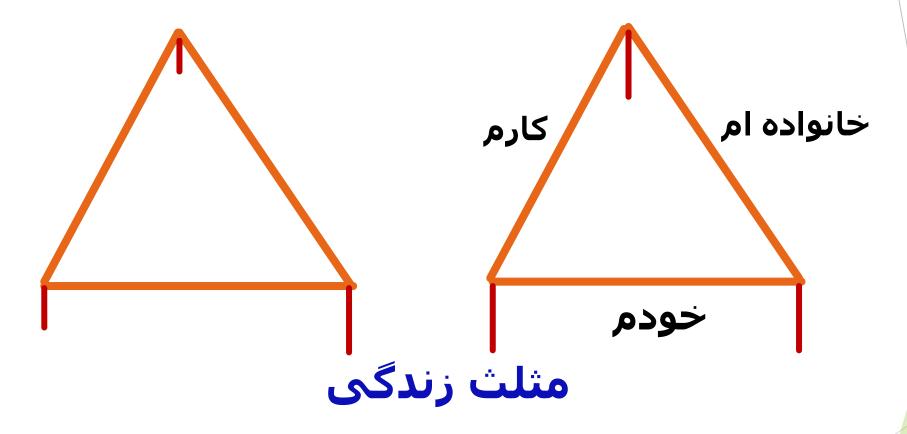
در سال ۱۳۷۰ که موسسه محک با هدف حمایت از کودکان مبتلا به سرطان با همت سعیده قدس و دیگر خیرین تاسیس شد، او به عنوان یکی از اولین متخصصان اطفال داوطلب (بدون دریافت حقوق) به این موسسه ملحق شد و بخش خون شناسی موسسه محک را راه اندازی کرد.

و در ادامه به عنوان رئیس هیئت امنای محک برگزیده شد و تا واپسین روزهای زندگی در این جایگاه به خدمت پرداخت.



پروفسور پروانه وثوق , مادر ترزای ایران





افراد بسیاری که به قله های موفقیت رسیده اند یک حسرت مشترک داِشته اند. بهای آن موفقیت , خانواده هایشان بوده است.





# امیرفرداد ظهیری

At 2mo if age presented with prolonged jaundice (cholestasis).

With imp.of BA, laparotomy was done, BDs were patent.

Liver Bx.: cirrhosis with extrahepatic obstruction.

He was treated with supportive therapy and followed.

# امیرفرداد ظهیری

#### FU:

Now he is 13 yr old, doing well, without any clinical symptoms of liver disease(compensated cirrhosis).

He is athletic, doing volleyball and football.

### Last PEx( 27/8/1403):

W/D ,W/N , no jaundice , ascites or edema. LT lobe of Liver is palpable , spleen 4-5 cm BCM.



امیرفرداد ۱۳٫ ساله (Biliary cirrhosis)

## محمد حاجی زادہ

A 3 mo old boy presented with cholestasis, suspected to BA, laparotomy was done, BDs were patent.

#### Liver Bx.:

1:Secondary biliary cirrhosis(17/4/1388)

2-Biliary cirrhosis (7/9/1390)

He was treated with UDCA and other supplements and followed.

## محمد حاجی زادہ

#### FU:

Now he is 15Yr old, without symptoms/signs of CLD.

**US:** Liver: NL., No splenomegaly or PHT.

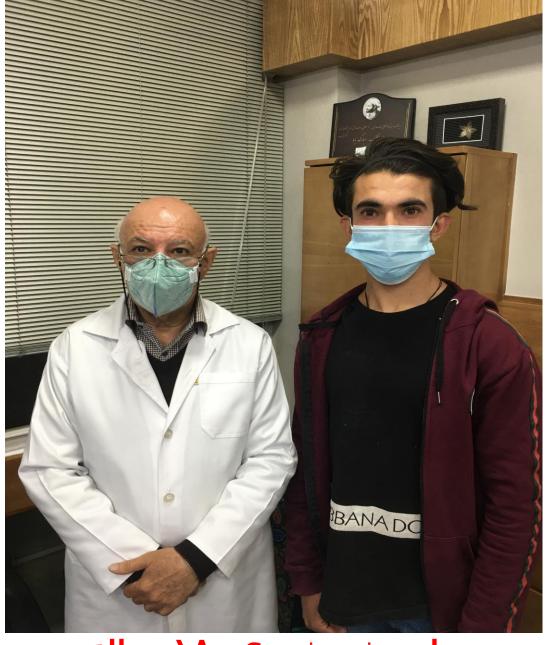
**Endoscopy:** No varices

CBC:WNL , ALT:20 , AST: 24 , PT/INR:NI





محمد حاجی زاده, ۱۵ ساله (Biliary cirrhosis)



علی شمشیری, ۱۸ ساله (BA)

## علی پزدانی

A10 yr old boy presented with yellowish skin and sclera for a few days and decreased LOC for 2 days (FHF).

PEx: He was deeply jaundiced with grade 2 encephalopathy.

No significant organomegaly or stigmata of CLD.

# ع.پزدانی

### Lab. Data:

ALT:325,125,67,72,PT:23/13, 18/13,

TB: 50, 42, 18.5, 14, DB: 24.6, 22, 11.7, 5.9

**CBC:WNL** 

UA: Blood 3\*, RBC:15-20, WBC:14-16, Bili:3\*

HAV IgM: Positive

# ع.یزدانی

Work-ups for other possible causes, mainly Wilson disease, AIH and drugs toxicity were negative.

With impression of FHF (AHF) with hemolysis due to HAV, was managed.

He also was candidate for Liver Tx.

## ع.یزدانی

He gradually improved and after two weeks was discharged with good condition.

He was followed in GI clinic regularly, without any treatment and had complete improvement.

#### FU:

Now he is 27 yr old, 6th yr medical student, completely normal and doing well.



عظیم یزدانی **, ۲۷ ساله** (HAV , FHF)

## زهرا.ش

A 40 days girl was admitted in GI ward with cholestasis.

At the time of admission she had jaundice, significant organomegaly, ascites and poor condition with significant bleeding tendency.

FHx. Was neg.(first baby).

All work-ups for possible etiologies were negative.

## زهرا.ش

Deu to clinical and lab. findings, with imp. of galactosemia, breast feeding was stopped and soy based formula was started.

Within a few days, gradually her condition improved.

Her jaundice decreased, bleeding tendency controlled, and LFT became better.

With final Dx.of galactosemia she was treated and followed.

## زهرا.ش

#### FU:

Now she is 26 yr old, completely normal, doing well.

No any sympt/signs of liver disease.

She is educated from high school successfully and recently married.





ز.ش , ۲۶ ساله (Galactosemia)

## ماجد قدری

A K/C of galactosemia from neonatal period.

He presented with cholestasis and significant hepatic failure.

He was treated with soy-based formula and followed.

FU: Now he is 19 yr old, completely normal, without any S/S of liver disease.

nto: 26 US: NL 14.4/8/10 ByR: 19 yr. wst. 60kg No Sympt. 18/21.

#### مجتمع درمانی شهید آیت الله مطهری SHIRAZ UNIVERSITY OF MEDICAL SCIENCES NEMAZEE HOSPITAL HISTORY & PROGRESS SHEET

Name (C 5 A	Unit No.
Name Last Last	Date 4.2.84 Service Locs
A 4 majo &, Known Car	2005
He had a literature	e of Galactosemia
in has no problem.	formula Al-110
P/E: Heart-lung - Abo	at all
1+ 01 ( )	$d \rightarrow Nl$ $wt = 5650$
It sided inguina	1 hernia.
Plan: 1. Dis Cont	in of ursodeony-vita
	K-
2. Multivia	tar 20 drop BID
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ماجد قدری ، ۱۹ ساله (Galactosemia)

### علی دشتیان

At the age of 40 days, was admitted in GI ward to bleeding tendency and elevated AST and ALT.

He was the first child of the family.

#### PEx:

He had jaundice without significant organomegaly. No more abnormal finding.

### علی دشتیان

#### Lab data:

CBC ,BS, BUN and other routine tests were normal.

```
AST:357 , ALT:310 , TB:6 , DB:3.7
```

PT:34 , INR:6 , PTT:47

Urine SA was positive.

Liver Bx.:Cirrhosis (cause biliary, such as PFIC)

### علی دشتیان

With impression of tyrosinemia, nitisinone (NTBC), 1mg/kg/day (5mg/day) was started.

Within a few days after treatment, he responded dramatically with correction of PT/INR and AST, ALT.

He was treated with NTBC (10 mg/day) and followed.

FU: Now he is 16yr old, completely normal and doing well.





على دشتيان, ۱۶ ساله (Tyrosinemia)

### Suchy liver disease in children

Hereditary tyrosinemia is a severe inborn error of metabolism, that can affect numerous organs, particularly liver, kidney and peripheral N.S.

In the first accounts patients, in 1950s, almost all died of liver disease in infancy or childhood.

Because early treatment is effective, tyrosinemia screening is increasingly induced in newborn screening Pannels around the world.

SA is a specific and sensitive marker for tyrosinemia, and is the preferred marker for newborn screening.

## عباس تنگستانی

A 6 yr old boy was referred to me with prolonged AST and ALT elevation.

A the that time he was apparently normal without any symptoms or signs of CLD.

Workups for possible causes were done and final Dx.was Wilson disease.

D- penicillamine and B6 were started and followed.

# عباس تنگستانی

After a few weeks, enzymes gradually decreased.

He was followed regularly and then referred to adult GI man.

#### FU:

Now he is 35 yr old, completely normal, without any problem, married and has one son.





عباس تنگستانی (۳۵ ساله) و فرزندش (Wilson Dx)



بهمن رستمی (۳۵ ساله, ویلسون)





Û



ف , ز , م , م مادر و نوه

### محمد علی پور حقیقی

A the age of 3 mo. was referred to me due to gallstone which was detected by US.

He was FT, without Hx. of any disease or medical therapy.

US was done for irritability, which detected 3-4 stones (8-10mm) in the GB.

PEx: was completely Nl.

### محمد علی پور حقیقی

His parents were very anxious and agitate about the method of his treatment including surgery and outcome.

He was treated with reassurance of the parents and followed without any medication and US every 6 mo in the first yr and then annually.

#### FU:

Now he is 19 yr old, without any symptom related to the stones.

•

## محمد علی پور حقیقی



شيراز، خيابان معدل، قبل از فلسطين(باغشاه)، ساختمان تابا كديستي: ٢١٣٢٧-١٤٤٠ تلفن: ٩٩٢۶٠٠٧٨٠٠ فكس ،٩٣٢٥٧٠٨٨

His last US |





محمد على پور حقيقى, ۱۹ ساله (Gallstone from infancy)

### **Gallstones**

#### Classification:

1-Cholesterol stones

2- Pigmented stones

3- silent (incidental) gallstone

## مريم جنگجوش

At the age of 5 mo. presented with irritability and poor feeding.

PEx.: Only had hepatomegaly.

Lab Data: Had significant AST and ALT elevation.

Liver Bx: PFIC

FU: Now she is 19 yr old, without significant clinical problem.

1+0) 10/44 PE: Ins organomegalf
No stigmate of cLD

No of Previous Shiraz University of Medical Science Admissions Nemazee Hospital HOSPITAL SUMMARY 127227 Date of Admission: Date of Discharge: Final DX: Hepatosphenomegaly finding cause (byler da)? Brief History: The Pt is a 5 mg of intent who was brought a cook inch. the hu of poor feeding the hu of tever 2-3 days ago are ha of cf Caugh - comiting rinnithen initability since 3 mg Positive Findings (Clinical): HC: 40cm L1:68 v.1:600gr +ve ha of liver da in her sister P/= : NO Positive findings (Laboratory , X-ray , EKG , etc): P.T: 13/3 Cont





مریم جنگجوش, ۱۹ ساله (PFIC)

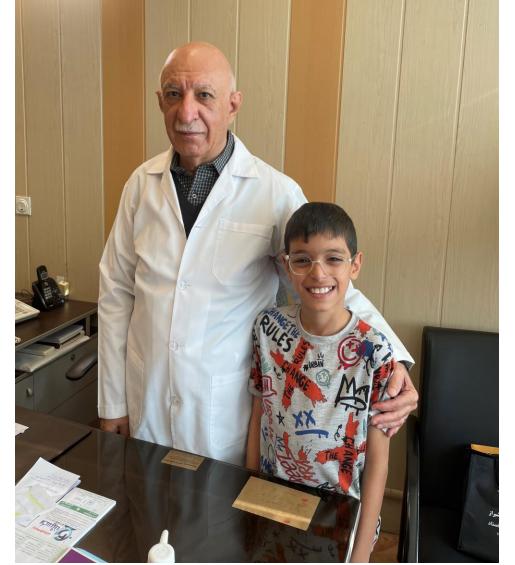
### محمدسرموري

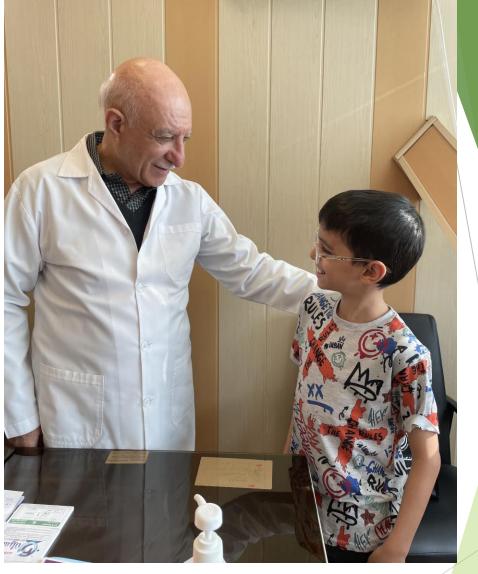
A the age of 12 days was admitted in NICU, with severe watery diarrhea and signs if severe dehydration.

Work-ups for all possible causes were done and finally with impression of GG malabsorption, Galactomine19 formula started and had complete improvement.

He was discharged with good condition and followed.

FU: Now he is 10 yr old, completely normal and doing well





محمد سرموری , ۱۰ ساله (GG Malabsorption)

#### مريم-ع

A 14 yr old girl with Hx. of GI symptoms from early life and with final Dx. of malrotation underwent surgery.

She had significant wt.loss after surgery.

She again developed abdominal pain and significant post-prandial vomiting and her wt. loss got worse.

She was admitted in our ward for Dx.and management.

مريم-ع

Our Dx.was cast (SMAS or Wilke, sSx).

With appropriate management, within a few days gradually, she tolerated feeding and after 10 days was discharge with a good condition.

#### FU:

After one mo., 3mo. and 6 mo. she was completely asymptomatic, and had significant wt. gain.





مریم-ع (CAST/SMAS)





م- عاجزپور (یک ماہ بعد از ترخیص)

### ابوالفظل سليماني

A \? yr old boy presented with Hx of severe RAP without more symptoms, for 2 days, from early life.

Intervals: were 2-3 mo.

FHx: was positive for migraine.

He was visited by many physicians, had frequent hospitalization and was treated with impression of abdominal migraine without improvement.





سلیمانی (Malrotation)

## محمد رامش

A 26 yr old young man, a K/C of FMF, from 5 yr of age.

HE was treated with medacin (colchicine), 1mg/day.

#### FU:

Now he is completely normal, married, has one child



محمد رامش (FMF)



فاطمه دشتی, ۱۸ ساله (FMF)

# **FMF**

1-How do you follow patients with FMF?

2-What is the most serious long term complication of this disease?



# غلامعلى نيكنام

An 11 yr old boy presented with Hx. of abdominal pain, diarrhea and significant wt.loss for about 2 yrs.

A the time of admission he was emaciated, chronically ill with severe FTT.

Work up for possible causes were done.

Final Dx. was CD.

After starting GFD, his symptoms improved within a few mo.s.

# غلامعلى نيكنام

## FU:

Now he is 44 yr old ,still on GFD, completely normal, without any symptom.

Married and has 3 children.



غلامعلی نیکنام و فرزندش (۴۴ ساله ,سلیاک)

# علی فارسی

A 5 yr old boy was referred to me with Hx.of CAP, flatulence, on and off loose stools and bad odor gas passing.

With imp. of CD workup was done and the findings were in favor of this disease.

**GFD** was started.

After a few mo. all of the symptoms improved.

FU: Now he is 28 yr old, still on GFD, doing well and is completely asymptomatic.





علی فارسی , ۲۸ ساله , سلیاک

# محمد باقرى

۱۸ ساله , وزن ۷۰ کیلو گرم

بدون علامت باليني

تست مثبت TTG,lgA در آزمایش روتین

تایید تشخیص سلیاک با بیوپسی روده

تحت درمان با رژیم فاقد گلوتن







ط.. معینی, ۶ ساله K/C of: DM , HPT and CD

## میکائیل رحیمی

A 5 yr old, K/C of CD.

Poorly controlled (poor compliance), with poor growth.



میکائیل رحیمی (CD)

# کوثر عسکری

A K/C of CF and CD.

She presented with diarrhea and malabsorption from early life and CF was diagnosed, treated with creon and other supportive therapy.

At age of 4 yr, she had increased BM, flatulence and significant wt.loss, despite adequate doses of creon.

Suspected to CD, work up was done and CD was diagnosed.

After starting GFD, her symptoms improved.

FU: Now she is 12 yr old with good condition.



کوثر عسکری, ۱۲ساله (CF and CD)





مهدی محمدی, ۱۸ ساله (CF)



رضا احمدی ۱۸٫ ساله (CF, from infancy)

# امیرمحمد احمدی

A 6 yr old boy presented with Hx of RAP, V/V(non-bilious) and diarrhea for 2-3 days for about 2 yrs.

Time of onset: Usually begun in the midnight

Intervals: 2mo.

Had frequent hospitalization, and with impression of food poisoning or viral GE, was discharged.

# امیرمحمد احمدی

He had positive Hx of motion (car) sickness.

FHx: was positive for migraine( mother)

PEx: was Nl. wt:18kg

With impression of CVS, propranolol 20mg/day started and followed.

# امیرمحمد احمدی

## FU:

He was treated with propranolol for 9 mo, and had complete improvement.

Propronolol was tapered to D/C and followed

Now he is 15yr, doing well, without any problem.





امیر محمد احمدی, ۱۵ ساله (CVS)

# ثمین اجرایی

A 4 yr old girl presented with Hx of recurrent hiccups without any more symptoms, for a few hrs, for about one yr.

Symptoms usually begun early morning.

Intervals: 2-3 mo.s

PEx: was Nl.

FHx.: Was positive for migraine (grand mother)

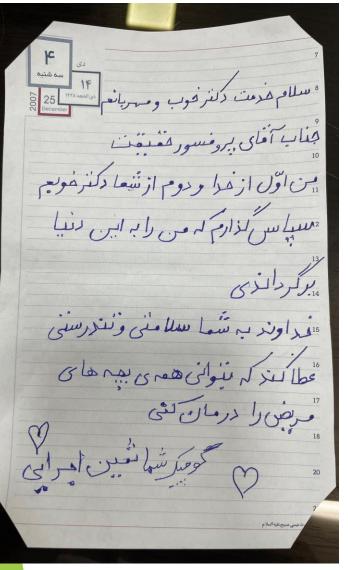
# ثمین اجرایی

With impression of CVS, propranolol was started and followed.

She had complete improvement and after 8 mo. of treatment, propranolol, tapered and was D/C.

### FU:

Now she is 8 yr old, doing well, without any problem.







ثمین اجرایی , ۸ ساله (CVS)

# آوا کهزادی

An 15 mo. girl presented with Hx.of RAP, N/V and diarrhea for 3-4 days, every 2 weeks for about 3-4 mo.

Attacks usually begun in the morning.

FHx was positive for migraine( grand mother).

PEx.: NI

# آوا کهزادی

With imp.of CVS, propranolol was started and followed.

## FU:

She improved completely and had no more symptoms during FU.



آوا کهزادی, ۲ ساله (CVS)



آرزو رحیمی،۱۶ ساله (CVS)

# **CVS**

- 1- CVS no longer considered to be rare in children or adults.
- 2- The prevalence of CVS in children is estimated at 1.9-2.3 %.
- 3- The average age of Dx. was 9.6 yrs ,while the average age of onset of symptoms was 5.3 yrs.

- 4 Has two types:
  - 1- pediatric onset 2- adult onset

# **CVS**

How do you:

1-Diagnose?

2- Treat?

and

3-What is the duration of treatment?

# World J Gastrenterology, 2007 Mar 28;13(12)

Cyclic vomiting syndrome in children: Experience with 181 cases from southern Iran

<u>Mahmood Haghighat</u>, <u>Seyed Mohammad</u> <u>Rafie</u>, <u>Seyed Mohsen Dehghani</u>, <u>Marzieh Nejabat</u>

#### **CONCLUSION:**

There is a significant lag time between the onset of clinical symptoms and the final diagnosis of CVS in our area.

In patients with typical clinical presentations of CVS, who are examined by an experienced physician, invasive workup is not necessary.

Propranolol appears more effective than amitriptyline for prophylactic use in children with CVS.

# Middle East J Dig.Dis. 2023 Jan;15(1):32-36.

Relapse Rate of Clinical Symptoms After Stopping
Treatment in Children with Cyclic Vomiting Syndrome

Mahmoud Haghighat <sup>1</sup>, Maryam Gholami Shahrebabak <sup>1</sup>, Seyed Mohsen Dehghani <sup>1</sup>, Maryam Ataollahi <sup>1</sup>, Nazanin Amin Farzaneh <sup>1</sup>, Samaneh Hamzeloo Hoseinabadi <sup>1</sup>, Hazhir Javaherizadeh <sup>2</sup>

### **Methods:**

Records of 504 patients below the age of 18 years with CVS who were treated with propranolol from March 2008 to March 2018 were reviewed.

Relapse Rate of Clinical Symptoms After Stopping

Treatment in Children with Cyclic Vomiting Syndrome

#### **Conclusion:**

The findings of this investigation show that the duration of treating CVS with propranolol could be shortened to 10 months with a low percent of symptoms relapse and this shortening may be effective in preventing the undesirable side effects of the drug.

## زهرا محمدی

An  $\Delta$  yr old girl referred for evaluation due to very high ALP levels which was detected on routine work up.

Past medical Hx.: was non-significant for any disease.

PEx: NL

# زهرا محمدی

## Lab data:

**CBC:WNL** 

ALT:57, AST:45, GGT:120

ALP: V۶9+ , 6950 (2 weeks later)

Ca, ph, BUN, Cr: NL

**US:NL** 

# زهرا محمدی

With impression of TH of infancy and early childhood, she was followed.

### FU:

**ALP** was checked at:

ALP: V۶9+ , 6950 (2 weeks later)

2mo:3475 , 4mo:2175 , 6mo:852



زهرا محمدی (TH)

### What is TH of infancy and early childhood?

### Is characterized by:

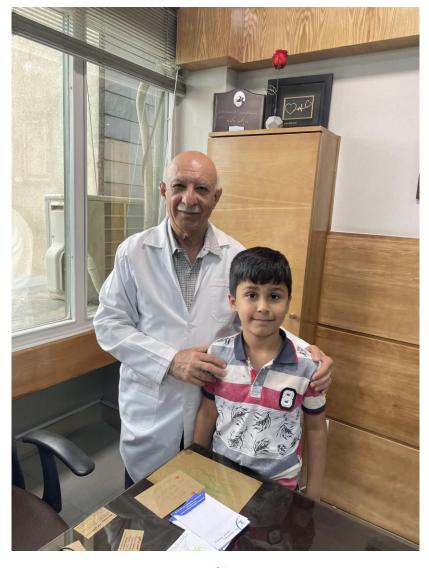
1-Marked elevation of ALP level

2- Absence of Hepatobiliary or bone disease

3- Return to normal within weeks or months

## TH of infancy and early childhood

- 4- Occurs mostly in infants and children younger than 5 yr, but can be see in older ages, even in adults.
- 5- ALP can elevate up to 20 times of pediatrics ULN and 50 times of adults ULN.
- 6- A second rise occurs in early puberty
- 7-ALP gradually return to normal within 2-3 mo, but may persists up to 6 mo.



رضا آزادی (TH)

### مهدی نصیری

A19 yr old boy with prolonged AST/ALT elevation, with impression of liver disease, was referred for evaluation and management.

There was no Hx.of any symptom in favor of liver disease.

PEx:W/D, W/N, no abnormal finding in favor of liver disease.

But had a significant specific abnormal finding....?

Cuff muscle hypertrophy

Dx: Myopathy

#### **Dr.Kamalian Ultrasound Clinic**

#### کلینیک سونوگر افی دکتر کمالیان

متخصص رادیولوژی سونوگرافی سی تی اسکن و ام آر آی

همکار گرامی: -با سلام و احترام سونوگرافی شکم:

کبد با اندازه و اکوی مناسب دیده شد .توده فضاگیر رویت نمی شود .سیستم صفراوی داخل و خارج کبدی دیلاته نمی باشد. کیسه صفرا بدون دیلاتاسیون و سنگ دیده می شود . ضخامت جدار کیس صفرا طبیعی است.

> طحال به سایز mm 106 و اکوی افزایش یافته رویت شد. بررسی از نظر بیماری زمینه ای (هموکروماتوز و متابولیک) توصیه می شود.

> > پانکراس در قسمت سر و بادی به سایز و اکوی مناسب رویت شد.

توده فضاگیر در آدرنال دو طرف رویت نمیشود.

اندازه کلیه راست <u>112 mm</u> و ضخامت پارانشیم آن <u>13 mm</u> است.

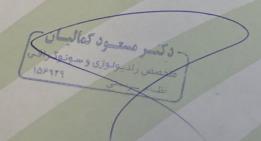
اندازه کلیه چپ 102 mm و ضخامت پارانشیم آن 16 mm است.

هر دو کلیه با اکو مناسب دیده می شود. توده فضاگیر مشهود نمی باشد.

شواهدی ازسنگ وهیدرونفروز رویت نمیشود.

مثانه ضخامت جدار مناسب دارد. سنگ و ضایعه فضاگیر درون مثانه دیده نمی شود.

مایع آزاد در حفره شکم رویت نمی شود.



تاریخ بذیرش: ۱٤٠٣/٠٧/٢٩ پزشک معالج: خانم د کتو فاطمه برامکی ش: ۲-۱٦٠٥ سن: ۱۸ سال .ه: آقای مهدی نصیری

		Hen	natology	
Test C.B.C.	Result	Unit	Normal Range	Differential
W.B.C.	₩4.1	10*3/μL	4.5 - 13	neutrophil (%)
R.B.Cimpal-F	5.44	10*6/μL	4.3 - 5.7	Lymphocytes (%)
Hgb	15.2	g/dL	13.2 - 17.3	Other WBC (%)
HCT	since 44.5 02	%	39 - 49	0 11.0. 11.2 0 (7.0)
M.C.V.		fL 11.0	Wint of Sales	V 1× 1
M.C.H.	27.9	pg	25 - 31	· Y 17.5
M.C.H.C.	34.2	g/dL	32 - 37	سه د افای مهادی لصیری
Platelets	176	10*3/μL	140 - 440	
RDW-CV	13.8		11.8 - 14.5	
PDW	11.2	fL	9.4 - 18.1	
MPV	9.6	fL	8.1 - 12.4	
P-LCR	22.6	%	10.7 - 45	

L Clyrigat-R				Bioch
Test Test	Result	Unit	Normal Range	Taking State of
Urea	35	mg/dL	7 - 42	
Creatinine	0.7	mg/dL	0.5 - 1.4	17.11.2
Total Bilirubin (venous blood)	₹ 3.4 P	mg/dL	0.2 - 1.2	C . 20
Capillary blood (sampling from the Refference: www.ncbi/gov/Direct Bilirubin	/m/Pubmed/3673 0.2	mg/dL	< 0.2	e 10 mg/ di.
	0.2			
Indirect Bilirubin	2 3.2	mg/dL	0.1 - 1.1	
S.G.O.T	\$ 208*	IU/L	8 - 33	
S.G.P.T	<b>★ 233*</b>	U/L	4 - 36	
Alkaline Phosphatase	187	IU/L	65 - 306	
* = Confirmed by Reneated Analysis				

Ciridia 1982 2884	0 . ñ.7	n di	0.14	Co
Test   Hirabia (v. 1981)	Result	Unit	Normal Range	
P.T. ( Prothrombin Time )	12.6	Sec	13.5	in the area
P.T. Activity	99	%		
I.N.R.	(1.01)			
P.T. Control Time	12.5_	Sec	12.5	
P.T.T.	21.9	Sec	Up to 36	
P.T. ( Prothro	mbin Time )			

ي توسط دستكاه اتوماتيك انجام ميكيرد



Address : 5th Alley - East Moadel st - Shiraz

Phone : (071)32302943-32306523-32348955-32333792

1403/08/01 - 17:27:53

کد ملي : 5140088721

کد سال - Male - پزشك : جناب آقاي دكتر محمود حقيقت (24873)

Ric	oche	om	ich	11
DIL	ALI II	will.	DL	4

Test	Result	Unit	Method	Reference
CPK	H * 3954	U/L	IFCC	35 - 174
SGOT/AST	H * 140	U/L	IFCC	<37
SGPT/ALT	H * 220	U/L	IFCC	<40

#### \*: Rechecked

#### **Laboratory Signature**

Print On 1403/08/01-19:07:47 By 36



#### Laboratory Director: Dr.M.Moha

تر محمود محمدی ص پاتولوژی تشریحی و بالینی

# على آبخو

A 9 yr old boy presented with prolonged AST and ALT elevation, admitted for workup and liver Bx.

All laboratory findings were in favor of Wilson disease.

D-Penicillamine and B6 were started and followed.

After 4 mo. of treatment AST, ALT became normal, but PT, INR still were prolong.

# على آبخو

We suspected to AIH, prednisolone and azathioprine were added to penicillamine.

After one mo. PT/INR became NI.

Liver Bx: was in favor of AIH.

Therefore with the Dx. of Wilson and AIH, he was treated concomitantly with D.penicillamine, Prednisolone and azathioprine.

# على آبخو

#### FU:

After a few mo. prednisolone was D/C and treated with D.penicillamine and azathioprine.

Now he is 17yr, without any clinical symptoms.

CBC, ALT, PT/INR were WNLs.





على أبخو (AlH and Wilson)

A 13 yr old girl transferred to our ward with FHF for liver Tx.

She had Hx.of behavioral Changes from 2-3 months ago.

She developed jaundice from 5 days and decreased level of consciousness from 2 days ago.

PMHx, Drug Hx and FHx: negative.

### P/Ex:

PR:145 , RR:24 ,T:37 , BP:100/80

Was icteric (deeply jaundiced)

No organomegaly, no stigmata of CLD.

Decreased LOC (G3 of encephalopathy)

### Lab. Data:

WBC: 7.2, 53.9, Hb: 9.9, 3.8, Plt: 275, 500

AST: 2170,1360 , ALT:1880, 980, ALP: 392, 315

TB: 39,73, DB: 28,36, INR: 2,2.4

### Lab.Data:

HAV IgM: Positive , COVID IgM: positive

ANA, ASMA, ALKA: Neg

Ceruloplasmin: NL , 24 hr urine CU: 371

Urine succinylacetone: Neg.

Brain MRI: NL

## Management

انیسه-ش- ب

She was managed for FHF.

Pre-transplantation workups also were done.

Since she had concomitant COVID infection, Liver Tx.was impossible.

Despite she was HAV positive, considering past Hx. and significant hemolytic anemia, with impression of Wilson disease, trientine and zinc were start.



### FU:

After starting treatment, she had significant improvement, clinically and para-clinically within a few days and was discharged with good condition.

She was followed at out patient clinic regularly.

- ► WBC:7.2 > 53.9 >39 >22 > 20 >16 > 7.3
- ► Hb:9.9 > 3.8 > 5.7 > 7.2 > 8.3 > 8.6 > 10.4
- ▶ Plt:275 >500 > 453 > 352 > 260 > 236 > 202> 116
- ► LDH: 6600 >1120 > 790 >711 > 598 > 570
- ► AST:2170 >1360 > 760 > 200 >100 >76 > 35
- ► ALT:1880 > 980 > 330 > 116 > 70 > 70 > 43
- ► Total bili: 39 > 73 > 40 > 18 > 19 > 15 > 5
- Direct bili :28 >36 > 20 > 11 > 9 > 8 > 2
- ► INR:2 > 2.4 > 1.8 > 1.4 > 1.3 > 1.2 > 1.12

### Suchy, liver disease in children, Fifth Edition

If the diagnosis of Wilson disease escapes detection, virtually all patients with ALF will die of hepatic or renal failure.

These patients never recover despite copper chelation therapy, plasmapheresis and require urgent Liver Tx.



زمان بستری



یک ماہ بعد از ترخیص





سه ماه بعد از ترخیص





شش ماه بعد

## هومن راه انجام

An 8 yr old boy, K/C of DM, referred to me with prolonged AST and ALT elevation.

At that time, he did not have any S/S of liver disease.

#### PEx:

Only had palpable liver. No stigmata of CLD.

## هومن راه انجام

Considering his underlying disease (DM), the first impressions were fatty liver versus AIH.

Workup for all possible causes including liver Bx. were done which were in favor of AIH.

Prednisolone and azathioprine (azaram) were started. After 24 hrs, he developed DKA.

Therefore pred. was D/C and azathioprine monotherapy continued.

### هومن راه انجام

After a few weeks, enzymes decreased and finally became normal.

#### FU:

Now he is 14 yr old, on 75 mg/day azaram, has good condition and LFT is WNL.

**Note:** to the best of our knowledge, he is the first case of AIH, who is treated with azathioprine monotherapy from the beginning of treatment.





هومن راه انجام (AlH, DM)

# آوینا رضایی

A K/C of AIH from 7 mo of life.

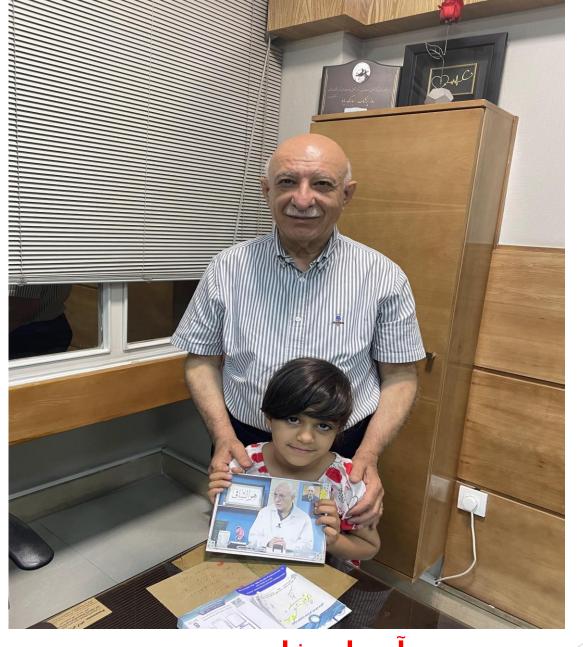
She was treated with prednisolone and azaram and followed.

Had complete improvement clinically and para-clinically.

After 3 yr of treatment, liver Bx was done and medications were D/C.

#### FU:

Now she is 6 yr old, completely normal, clinically and Para-clinically.



آوینا رضایی (AlH)







امیر ساده ۱۷ ساله (AIH)



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م.رحیمی, ۱۹ ساله (AlH)

## زهرا-ط

A 7yr old girl presented with jaundice for about one mo.

She had HAV hepatitis(HAV-IgM positive) at the beginning of disease.

Past Hx. and FHx.were negative.

### PEx.:

She was jaundiced, had firm hepatomegaly(3-4 cm.BCM) No stigmata of CLD.

زهرا-ط

Lab. Data:

CBC: NL

ALT:86 , AST:97 , ALP:1260 , TP:7.9 , Alb:5

TB:3.7, DB:1 PT:13, INR:1

US: Except hepatomegaly, no more abnormal finding.

#### Liver Bx.:

Destructive cholangiopathy with fibrosis (drug induced or PBC)

زهرا-ط

There was no Hx.of using any medication.

With Dx.of PBC, UDCA was started and was followed.

#### FU:

Two mo.Later: No clinical symptoms , PEx:NI , ALT:12

After one yr of treatment with UDCA, she is completely normal with normal LFT.

### Considerable points in our case

1-She is the first case of PBC reported in children from IRAN.

2-She is one of the youngest case of PBC reported in children worldwide up to now.

3- She had negative AMA.

4- Most probably her disease was triggered by HAV hepatitis.





زهرا - ط ۷٫ ساله (PBC)

### محمد حیاتی

A 6yr old boy presented with abdominal pain, anorexia, pallor and weakness for one mo. and recently was unable to walk.

He was visited by a few physicians, received different treatment, but had no improvement.

He was referred for GI evaluation (endoscopy).

#### PEx:

He was pale, sick looking and chronically ill. Liver was palpable with tenderness. Heart sounds were muffled.

### محمد حیاتی

With impression of heart problem, was referred to a cardiologist.

He had severe dilated cardiomyopathy with 20% EF.

He was admitted in cardiology ward for management.

FU: Despite extensive and appropriate management, unfortunately he died.



A 28 mo.old girl presented with Hx, of abdominal distention, non-bilious vomiting, which was more significant after feeding and also constipation from early life.

She had repeated hospital admissions and with impression of HSP, Bar. enema and rectal Bx. were done which were NI.

•

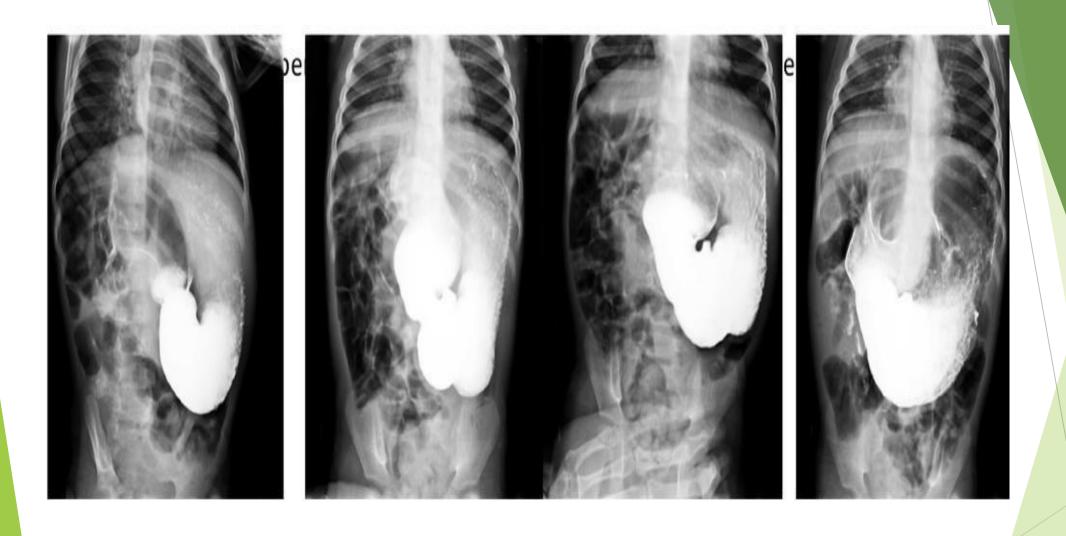
She was admitted in our GI ward.

PEx: She was imatiated and chronically ill.

Our impression was:

upper GI obstruction

Therefore UGS was requested which showed.



# **UGIS**

#### **UGI Report:**

Significant gastric distention with delayed gastric emptying .

No contrast passage to the duodenum.

For better evaluation, of congenital anomalies such as duodenal web, endoscopy was done.

#### **Endoscopic findings:**

Severely distended stomach with food residue.

Pyloric canal was stenotic, pyloric orifice was not seen, and snare could not bee passed to the bulb.

She was operated, Post op Dx.was:

Preduodenal portal vein(PDPV).

She had complete improvement after surgery.

PDPV is a rare congenital vascular anomaly, in which the portal vein is located anterior to the duodenum instead of posterior as in the normal anatomy



A 3 mo.infant, CC: poor Wt gain





امیر علی مومنی, ۲۲ ساله

(NL,FU)



رضا و یسنا پارسامهر (CD)





ع.عسکری ۱۸٫ ساله

IBD(UC) and PSC

Controlled on mesalamine and UDCA

#### اسماعیل خلیلی

12 yr old, K/C of DM, with short stature.

Wt: 20 kg

PEx:

Significant hepatomegaly

Lab.Data:

High FBS and increased AST/ALT levels

What is the Dx?

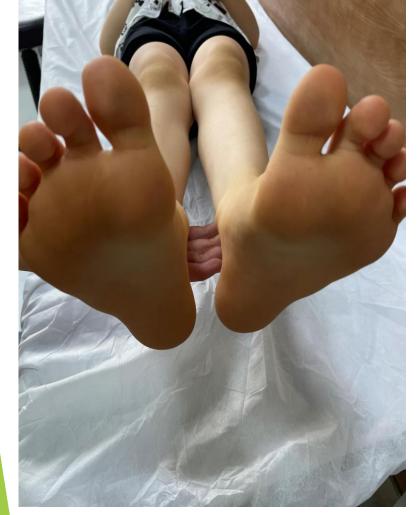
Mauriac Sx.



اسماعیل خلیلی Mauriac Sx.



س.روستایی: ۱۳ ساله با یبوست مزمن و اختلال رشد شدید Dx: HSD







What is the Dx.? What is the cause?







علیرضا (۱۶ ساله) و زهرا( ۱۲ ساله ) مقداری

(Cryptogenic cirrhosis)



140/19/19 Byp, 1641. W/ WILLY CBC, WILL
NO Sympt. Pod / OLS ELR : 5

Plan: Mesaline 1500 9, d. Stort, wen

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LF-Y/Y/V NO Sympt. W/+74/19 00000 CR12 wey

PE1 ok. CBC2 conta CR12 wey

Stool Colproduct = 18.7 plans Chonoscopy. Obsess in the second CC: bose start + Blood. PE/No don't finding

CBC: NNL. Start Calprot. = 22.7 Plan Repr te Dr. 166 ( 1989) file:///C:/Users/app7.NGH/AppData/Local/Temp/DMODTemp/DMODTmpA6EE4.xml 09/05/2020



A 11yr old boy presented with pruritus and yellowish skin and sclera for about 2 mo.

#### PEx:

He had jaundice with palpable liver.

No stigmata of CLD or any abnormal finding.

Past Hx and FHx: negative

All workups for possible underlying diseases were negative.

#### Lab.data:

**CBC:Nl** 

ALT:96, GGT:45, TB:13.8, DB:3.2, TP:7, Alb:4.3

**MRCP:NI** 

Liver Bx: Acute hepatitis

#### Management:

UDCA and rifampin started and followed.

After1 and 2 mo. of treatment, had no significant improvement.

Liver Bx. was repeated:

Chronic hepatitis (G:11/18, S:1/6)

Suspected to AIH

With imp.of AIH, prednisolone and MMF were started.

After 2 mo. of treatment, jaundice persisted.

Lab data:

CBC:NL , PT/INR:NL , ALT:275, TB:13.9 , DB:1.5

Prednisolone, MMF and UDCA continued for 2 mo.s. Jaundice persisted.

PEx: Had jaundice with palpable liver.

Lab data: CBC, PT/INR:NL, ALT:310, TB:9, BD:1.3

Phenobarbital was added to Prednisolone, MMF and UDCA.

After a few mo. of treatment, he had significant improvement.

Prednisolone tapered up to 5mg/day, phenobarbital was D/C and other medications continued.

#### FU:

After about 2 yrs of treatment, now he is completely normal, doing well, without any S/S if liver disease.

#### Last lab data:

CBC, PT/INR:NL, TB:2, ALT:137

#### **Medications:**

Prednisolone: 5mg, MMF: 2GM, UDCA: 900, daily





ایلیا فرازمند



ایلیا فرازمند

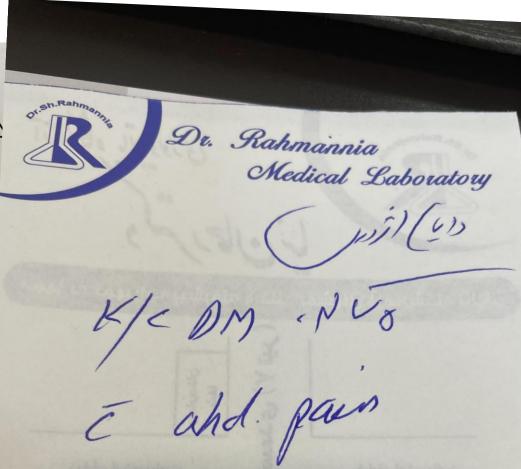
## SPIRAL CT SCAN CONTRAST:

Multiple axial im and oral contrast a Normal size liver. found.

Gallbladder is norn Spleen and pancrel Both adrenal gland Both kidneys are n of hydronephrosis. No paraaortic lymurinary bladder at The hallow viscus No sign of ascites There is a large inbout 70 x 48 x 9 and cecum. No en liagnosis is a more spleen and secum.

1GH





العین العین

تاريخ: 1403/01/29

نام بیمار : دایان اژدری

با سلام

همكار محترم :

#### Sonography:

in size and echopattern with no signs of diffuse liver disease, l cystic or solid liver lesion. Portal and hepatic veins are normal

no distension, stone or wall thickening or any signs of GB and intrahepatic biliary ducts are not dilated.

ographically, in size and echotexture.

in size with no stone, mass or hydronephrosis or evidence of ical thickness and echotexture are normal bilaterally. Ureters are

ortic regions are intact with no evidence of pancreatic enlargement pathy.

in upper or lower abdomen with no any solid mass lesion but there optated large hypoechoic cystic area as RT abdomen about which the possibility of mesenteric cyst should be ruled out. No vidence of inflammatory or malignant changes are detectable. al with no evidence of distention, interloop lesion or significant g however if clinically is suspicious of enterocolitis, further sted. No mesenteric adenitis or any evidence of retroperitoneal e seen too.

kable with no stone, mass, wall thickening or other sings of rgans are unremarkable with no mass or adenopathy.