La gestione delle epatopatie pediatriche quando diventano adulte

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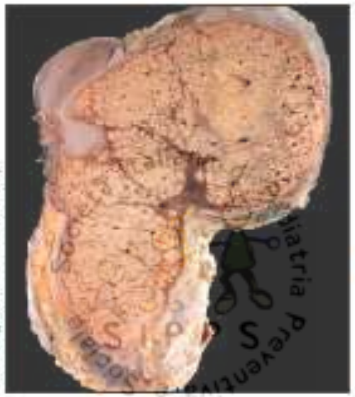
Liver disease in Europe



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About 29 million people in the European Union have chronic liver disease according to *The burden of liver disease in Europe: A review of available epidemiological data*, published recently in the *Journal of Hepatology*. Alcohol consumption, viral hepatitis B and C, and metabolic syndromes linked to overweight and obesity are reported to be leading causes of liver cirrhosis and primary liver tumours.

Liver cirrhosis is responsible for around 170 000 deaths in Europe annually, with wide variations between countries—ranging from about one per 100 000 Greek women to 103 per 100 000 Hungarian men dying each year. About 90% of individuals in Europe infected by viral hepatitis are not aware of their status, which is of great concern: chronic hepatitis B affects 0·5–0·7% of the European population, and the prevalence of chronic hepatitis C was 0·13–3·26% in the past decade. Moreover, the prevalence of non-alcoholic fatty liver disease is 2–44% in the European population. In terms of alcohol consumption, Europe is the region of the world where the heaviest drinking occurs; in some European countries, the mortality rate from alcohol-related liver diseases is as high as 47 per 100 000 inhabitants. A recent study done in Canada showed that a rise in alcohol prices of 10% was associated with a reduction in alcohol-related deaths by almost 32%, which could provide impetus for alcohol-control policies in Europe.

The substantial burden of liver disease-associated mortality in Europe means that governments and health-care providers must tackle liver disease in a much more proactive fashion—taking a cue from diseases such as breast cancer. A higher public health priority and use of non-invasive tests to screen for early stages of fibrosis are required. There is an urgent need to implement prevention programmes, and research will be needed to develop novel treatments to address the problem. The European Parliament's meeting on the burden of liver disease in Europe on Feb 20 in Brussels should be a first step towards greater care for the health of Europe's livers. ■ *The Lancet*

For ***The burden of liver disease in Europe: a review of available epidemiological data*** see http://dx.doi.org/10.1016/j.jhep.2012.12.005

For the **Canadian study** see http://onlinelibrary.wiley.com/doi/10.1111/add.12139/abstract

Hepatology
WILEY
HEPATOLOGY 2009;49:880-886

LIVER FAILURE/CIRRHOSIS/PORTAL HYPERTENSION

Graft Fibrosis After Pediatric Liver Transplantation: Ten Years of Follow-up

Rene Scheenstra,[1,2] Paul M.G.J. Peeters,[2,3] Henkjan J. Verkade,[1,2] and Annette S. H. Gouw[2,4]

In this study we have shown a profound increase in the **prevalence of fibrosis** in pediatric grafts from 31% at 1 year after transplantation to almost **70% after 10 years**, and an increase in the severity of fibrosis with a progression to severe fibrosis in more than 25% of the grafts after 10 years. Transplant-associated factors are related to the development of fibrosis, and even to the development of “late” fibrosis (that is, starting after the first year)

We attempted to identify the (severity) of fibrosis using biochemical liver functions…… However, most of the liver tests were only mildly elevated, and **even normal in most individual cases.**

1 Prevenzione

2 Ricerca

2008, Volume 371, Number 9628,

VENTO S, and NOBILI V

Aminotransferases as predictors of mortality

“the finding of **even modest increases of aminotransferase concentrations should not be overlooked**, as it could provide an opportunity for clinicians to reveal and treat not only serious hepatic diseases but also to modify detrimental habits that can be life-threatening in the long term.”

Special Review

EASL EUROPEAN ASSOCIATION FOR THE STUDY OF THE LIVER | JOURNAL OF HEPATOLOGY

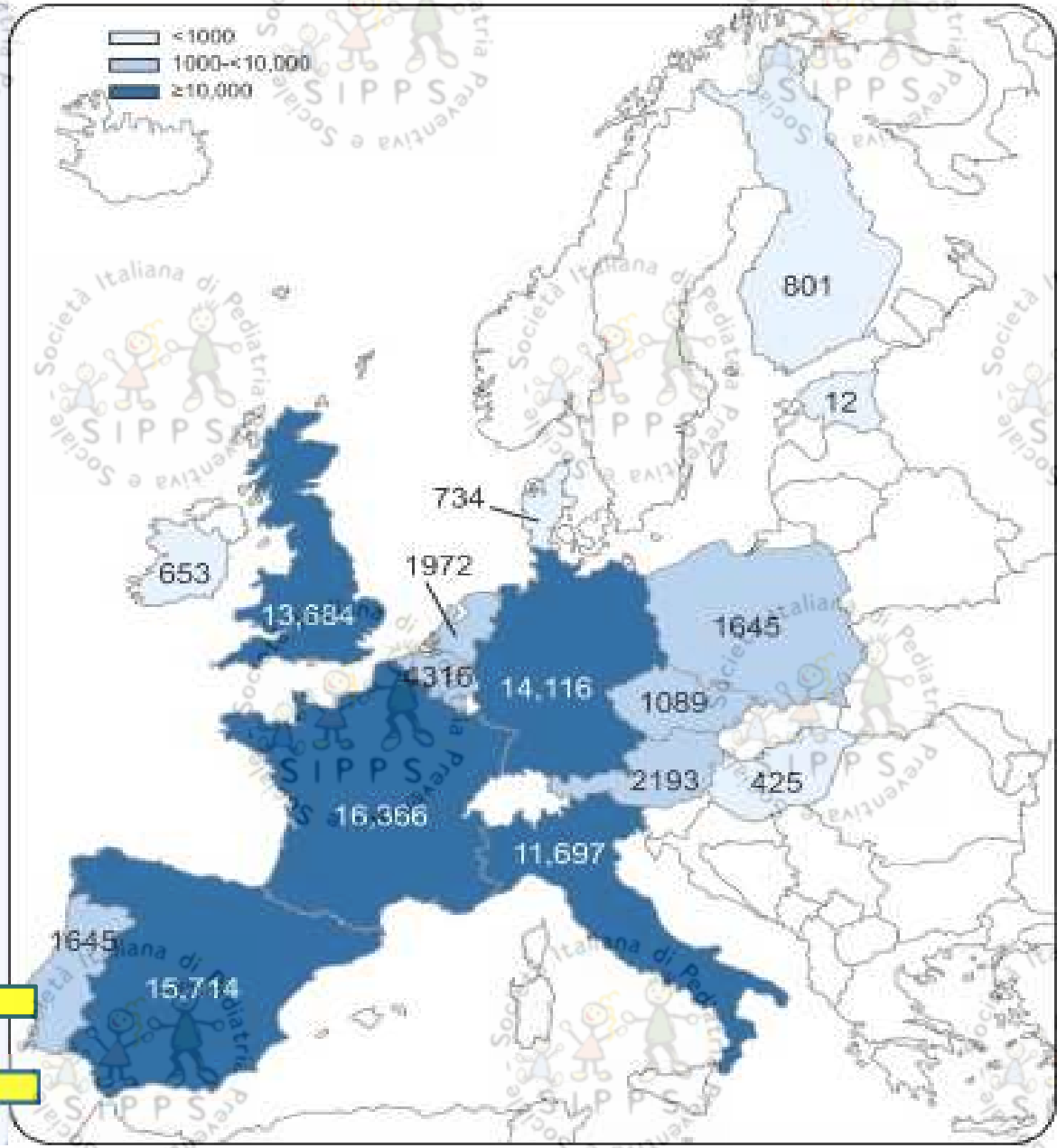
The burden of liver disease in Europe: A review of available epidemiological data

Martin Blachier[1], Henri Leleu[1], Markus Peck-Radosavljevic[2,*], Dominique-Charles Valla[3], Françoise Roudot-Thoraval[1,*]

[1]*Department of Public Health, Hôpital Henri Mondor, Université Paris-Est Créteil, France;* [2]*Dept. of Gastroenterology & Hepatology, Medizinische Universität Wien, Währinger Gürtel 18-20, A-1090 Vienna, Austria;* [3]*Service d'hépatologie, Hôpital Beaujon, AP-HP, Université Paris Diderot and INSERM U773, Clichy-la-Garenne, France*

Key Points

- A review of 260 epidemiological studies published in the last five years
- Liver cirrhosis is responsible for around 170,000 deaths in Europe each year, with large inter-country variation
- Liver cancer is responsible for around 47,000 deaths per year in the EU
- More than 5500 liver transplants are performed in Europe per year
- In some European countries the mortality rate from alcohol-related liver disease is as high as 47 per 100,000 inhabitants
- 0.5-0.7% of the European population is affected by chronic hepatitis B
- The overall prevalence of hepatitis C in Europe is estimated at 0.13-3.26%
- The prevalence of non-alcoholic fatty liver disease (NAFLD) is 2-44% in the general European population (including obese children) and 42.6-69.5% in people with type 2 diabetes. There is the potential for this condition to become a serious problem in light of the obesity epidemic
- All of the major causes of liver disease are amenable to prevention and treatment
- Strategies are urgently required to reduce the burden of liver disease in Europe



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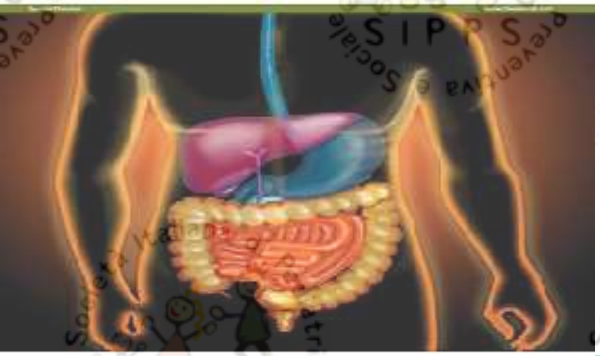

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Review
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Long-term challenges and perspectives of pre-adolescent liver disease

Nedim Hadžić, Ulrich Baumann, Pat McKiernan, Valerie McLin, Valerio Nobili

Chronic liver disease is a growing problem that has substantial effects on public health. Many paediatric liver conditions are precursors of adult chronic liver disease, cirrhosis, and hepatocellular carcinoma. Clinical management of Wilson's disease, autoimmune liver disease, and chronic biliary disorders, such as biliary atresia, which remains the most common paediatric chronic liver disease and indication for liver transplantation, is similar in children and adults. In the past 10 or so years, paediatric hepatology has expanded into neighbouring clinical areas, such as metabolic liver diseases and systemic conditions with liver involvement. In this Review, we aim to describe some of these disorders, and outline their natural history and possible differences between management in adults and children to stimulate further debate on the optimal transition of care between paediatric and adult specialists.

Lancet Gastroenterol Hepatol 2017

Paediatric Centre for Hepatology, Gastroenterology and Nutrition, King's College Hospital, London, UK (Prof N Hadžić MD); Paediatric Gastroenterology and Hepatology, Hannover Medical School, Hannover, Germany

	Specific clinical findings	Medical management
Biliary atresia	Established chronic liver disease, possible education difficulties in patients who have not received a transplant	Monitor and treat complications of portal hypertension and cholangitis, preferential status on transplant waiting lists might be necessary
α1 antitrypsin PiZ deficiency	Variable degrees of chronic liver disease	Involve adult respiratory physicians in monitoring of respiratory function
Progressive familial intrahepatic cholestasis	Possible relation to development of gallstones, cholestasis of pregnancy, and drug-induced liver injury	Choleretics and effective control of pruritus, including biliary diversion options
Cystic-fibrosis-related liver disease	Progressive portal hypertension, exocrine and endocrine pancreatic insufficiency	Probable psychological and social difficulties, consider for liver transplantation only in absence of very advanced lung disease
After liver transplantation	Drug non-adherence common, graft remodelling can induce secondary portal hypertension	Overall treatment similar to adults; monitor for nephrotoxicity
Non-alcoholic fatty liver disease	Large-scale epidemiological problem, end-stage liver disease exceptional in adolescence	Lifestyle modifications when possible, monitor for diabetes and cardiovascular complications
Autoimmune liver disease	Many patients on maintenance low-dose immunosuppression, frequent problems with adherence	Consider careful weaning or modification of immunosuppression ahead of pregnancy
Chronic viral hepatitis	Most patients have near-normal biochemistry and mild histological changes	New interferon-free regimens need to be approved for treatment of hepatitis C virus infection in children and adolescents
Wilson's disease	Neurological involvement less common in children and adolescents	Overall treatment similar to adults

Table 3: Specific features of chronic liver disease in adolescence

	Locus	Gene	Defective polypeptide	Extrahepatic features
Progressive familial intrahepatic cholestasis type 1 disease	18q21.31	*ATP8B1*	Aminophospholipid translocase FIC1	Affects gut, kidney, and lungs; associated with deafness
Bile salt export pump deficiency (familial intrahepatic cholestasis type 2)	2q24	*ABCB11*	Bile salt export pump	No
TJP2 deficiency	9q13–q21	*TJP2*	TJP2	Affects lungs and gut; associated with deafness
Arthrogryposis renal dysfunction cholestasis syndrome	15q26.1	*VPS33B*	VIPAR protein	Affects kidney and bones
Transaldolase deficiency	11p15.5–p15.4	*TALDO1*	TALDO	Affects skin and heart; associated with facial dysmorphism
MDR3 deficiency (progressive familial intrahepatic cholestasis type 3)	7q21.1	*ABCB4*	MDR3	No
Neonatal ichthyosis-sclerosing cholangitis syndrome	3q28–q29	*CLDN1*	Claudin 1	Affects skin and leucocytes
Neonatal sclerosing cholangitis	6p22.1	*DCDC2*	DCDC2	Associated with dyslexia, nephronophthisis, and deafness

Table 1: **Progressive familial intrahepatic cholestasis associated with low or normal serum γ glutamyl transferase concentrations**

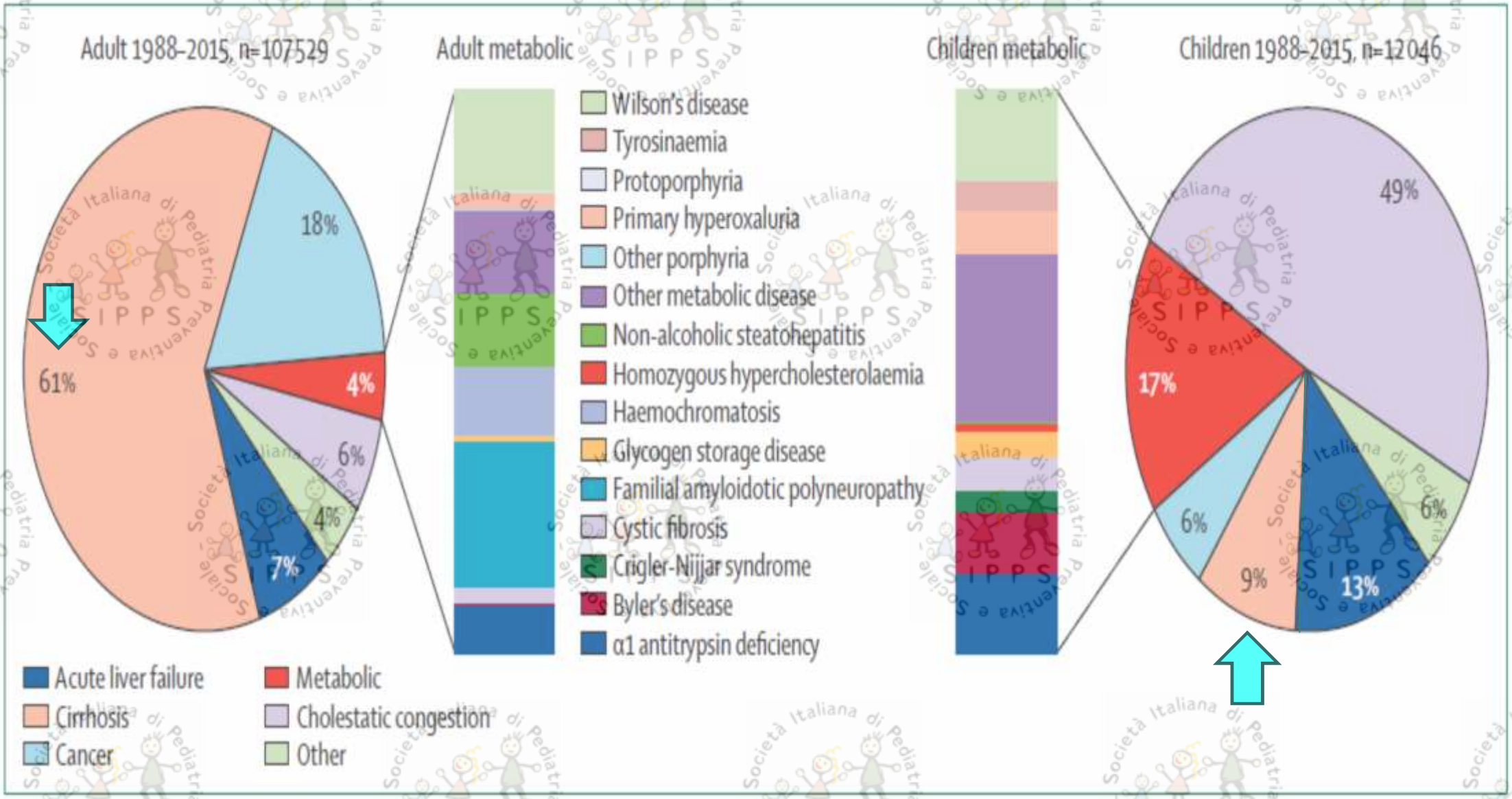

Figure: Differences in indications for liver transplantation between adults and children

Patients with chronic liver disorders who are leaving the paediatric age group continue to have substantial medical problems, which are complicated by individual behavioural, social, and educational difficulties. A new subspecialty—transitional medicine—is slowly developing; it is faced with the difficult task of offering a similar level of medical care with very different individual responsibilities. Health services worldwide are generally inadequately prepared for this major change, and hepatology is no exception. The complex needs of these fragile young people need to be recognised, and new services, such as adolescent psychology support services, need to be developed. Many of these patients have insufficient knowledge about their chronic conditions, because for most of their childhood their medical care has been coordinated by their guardians, typically their parents. The transition from this sheltered position to being a potentially vulnerable young adult who is inadequately prepared for the individual responsibilities, could have pronounced consequences, such as intermittent or total non-adherence to prescribed drugs or lifestyle, or complete dissociation from established medical follow-up.

Transitional Medicine

Parents

Non-adherence

Medical professionals need to acknowledge and respond to these new trends. Older paediatric patients should be educated, and active participation in their care should be promoted by paediatric hepatologists as they approach adolescence. This process could be formalised through pre-transition and transition clinics equipped to address the psychological and social needs of these emerging patients. Furthermore, referral pathways and the adult hepatologists taking over the formerly paediatric patients should be clearly identified. Adult hepatologists will need additional training in some specific paediatric disorders, such as biliary atresia and metabolic disorders. Finally, national liver transplantation programmes need to consider whether young individuals with end-stage chronic liver disease should be offered preferential status on the waiting lists. The conventional liver transplantation criteria were not designed to capture the complexities of care for this vulnerable new population, who compete for organs with adults. This situation could result in increased morbidity and mortality on waiting lists and massive disappointment for their families, past paediatric caregivers, and health-system economics.

Pre-transition and transition clinic

Referral pathways

Additional training

Liver Diseases: Children:Written question - 49864

Q Asked by **Mr George Howarth** (Knowsley) Asked on: 24 October 2016

Department of Health © 49864

Liver Diseases: Children

To ask the Secretary of State for Health, what assessment he has made of the implications for the health of children with liver disease of continuity of care as they make the transition to adult services.

A Answered by: **David Mowat**

The transition from children's to adults' health services is a matter for National Health Service providers and commissioners. To support effective transition commissioners and providers should be drawing on best practice and guidance such as the National Institute for Health and Care Excellence's guideline, Transition from children's to adults' services for young people using health or social care services which was published in February 2016.

How to do it: Key elements of transition policy

- Identification of adult centre and consultant
- An early start
- A written transition policy
- A flexible policy on timing of events
- An education programme
- Opportunities for the young person to meet the adult team

Services
Centre for Rare Diseases
Liver
Transition clinic for patients with liver disease
Meet the team

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You are here: Home > Services > Centre for Rare Diseases > Liver > Transition clinic for patients with liver disease

Date: 11 April 2017 Time: 07:22

Transition clinic for patients with liver disease

The liver is a vital organ. Without it we would not be able to live. Apart from the brain, it is the most complex organ in the body. It has a wide range of functions and acts very much like a factory. One of its most amazing features is its regenerative power.

Our service sees patients who have had liver disease in childhood and now need to be seen in adult services. Many of our patients will have had a liver transplant as children

How to contact the service

Karen Pears – Medical Secretary

Email: Karen.Pears@uhb.nhs.uk

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New transition app to help young liver disease patients

02/12/16

A specialist research team at Coventry University has enabled Children's Liver Disease Foundation to launch an app which will help young people with liver disease to cope with the transition from paediatric to adult health services.

My Liver is designed to help young people have all the knowledge they need about their liver, their transition and their care.

Transition can come at a difficult time, so the app contains features to support independence in adult services, such as information on liver diseases, important contact details and a reminder feature to aid with medication and hospital visits.

Visit our Focus website to find out more and download the My Liver App.

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iLiver

EASL EUROPEAN ASSOCIATION FOR THE STUDY OF THE LIVER

ILIVER CONTRIBUTORS
Paediatrics Liver Disease
Prof. Valerio Nobili

Clinics and Research in Hepatology and Gastroenterology (2014) 38, 277–283

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MINI REVIEW

Tips and hints for the transition: What adult hepatologists should know when accept teens with a pediatric hepatobiliary disease

Salvatore Guercio Nuzio[a], Sarah Ann Tizzard[b], Pietro Vajro[c,*]

- Between 15 and 20 years
- Earlier = staff unaware of developmental issues
- Completion of school is a possible benchmark
- After achievement of "health literacy":
  - Understanding of their disease
  - Management independent of parental figures

- Educational Sessions:
  - disease characteristics
  - origin of symptoms
  - how to contact health professionals
- Instruction on:
  - treatment rationale & therapies schedule
  - dosing regimen & routes of administration
  - setting up medical appointments
- Training for self-management of medical regimen
- Start individual meetings with providers
- Use of modern technologies (support groups; text messaging; online health management)
- Offer adolescent friendly environment

- A joint structure (pediatric & adult staff) to facilitate dialogue & navigation of medical bureaucracy
- Presence of a "coordinator" [e.g. a nurse], mediating transitional organization & patient management
- Administrative and secretarial support
- Co-management & communication between primary care physicians / medical specialists
- Primary care physician & hepatologist fully informed on characteristics and complications of the disease
- Contrast teens resistance to leave pediatric care & pediatricians' reluctance to discharge grown patients

Table 1 Literature evidences on long-term follow-up of patients with pediatric onset chronic hepatobiliary diseases.

Disease (Reference)	No of patients	Mean age at Dx (range)	Mean age at last FU	Mean FU duration (range)	Late complications	Living without LTx	Living with LTx	Death	Notes
Biliary atresia Post-Kasai [4]	80	63 days (33–159 days)	24.7 years (20–31.9 years)	NA	By age 20, liver cirrhosis in half of the adult survivors > 20 years of age: 37% and 17% of the adult patients = cholangitis and gastrointestinal bleeding, respectively	5, 10, and 20-year survival rates of patients with their native livers = 63%, 54%, 44%	5 underwent living related LTx before age of 20	20% adults died of liver failure/LTx in their twenties	5 female patients gave birth to 9 children; 1 male patient fathered a child
Autoimmune hepatitis [5]	33	12.9 years (2.7–18.1 years)	NA	73 months (11–344 months)	Cirrhosis in 55% at diagnosis	Cirrhotic patients at baseline had a 10 years survival of 85% (range = 70–100%) comparable to non-cirrhotic patients: 75% (49–100%) ($P = 0.97$)	Of the 18 patients with cirrhosis, 5 had LTx	6 patients died and in 4 of them it happened following complications after LTx	Overall survival was significantly lower than the expected in the USA population
Primary sclerosing cholangitis [7]	52	13.8 ± 4.2 years (1.5–19.6 years)	NA	6.6 ± 4.4 years (0.2–16.7 years)	81% had concomitant IBD, lower plts, splenomegaly, and older age were associated with shorter survival Overlapping AIH (35% of children) or medical therapy did not affect survival	LTx free median survival = 12.7 years	11 children underwent LTx	1 child died	Compared with USA population, survival was shorter in children with PSC ($P < .001$)
Hepatitis B [8]	99 – 91 HBeAg [+], – 8 HBeAg [–]	HBeAg [+]: 7.4 ± 4.7 years HBeAg [–]: 4.8 ± 3.2 years	26.4 ± 5.5 years	14.5 ± 6.1 years after HBeAg clearance, 17.0 ± 5.1 years in the 8 baseline HBeAg–	89 = HBeAg seroconversion after 5.2 ± 4.0 years 4/89 = reactivation with cirrhosis 85/89: 4 HBeAg [–] hepatitis, 81 inactive carrier; 64 were untreated, 21 previously treated with IFN or other drugs Of the 8 HBeAg [–], 2 were HBeAg [–] hepatitis, 6 carriers	Of the 4 with baseline cirrhosis, 2 had HCC and remained alive and 2 lost the histological features of cirrhosis	NA	NA	Two patients with HBeAg [–] hepatitis and 1 with cirrhosis had experienced drug abuse
Hepatitis C [10]	224	5.8 ± 3.7 years	NA	6.2 ± 4.7 years (1–17.5 years)	Of 200, 6% achieved sustained viremia clearance and normalization of the ALT level Mean fibrosis score: 1.5 ± 1.3 for < 15 years of age and 2.3 ± 1.2 for ≥ 15 years of age	NA	NA	NA	

Table 1 (*Continued*)

Disease (Reference)	No of patients	Mean age at Dx (range)	Mean age at last FU	Mean FU duration (range)	Late complications	Living without LTx	Living with LTx	Death	Notes
NAFLD [12]	66	13.9 ± 3.9 years (3.2–19.6 years)	NA	6.4 ± 4.5 years (0.05–20 years)	4 patients with baseline normal fasting glucose developed type 2 diabetes 4–11 years after NAFLD diagnosis 13 liver biopsies in 5 patients over a mean of 41.4 (SD 28.8) months showed progression of fibrosis stage in 4 patients	NA	2 NAFLD recurred. One developed cirrhosis and needed re-LTx	2 patients died for decompensated cirrhosis	Survival free of LTx significantly shorter vs general population
Wilson disease [14]	229 – 61% hepatic, – 27% neurol, – 10% screened	21.2 ± 12.0 years (4–58 years for hepatic presentation)	NA	11.8 ± 8.6 y (hepatic presentation)	84% cirrhotic patients survived 20 years after diagnosis	35% stabilized or improved on chelation (26% fully recovered, 24% improved)	8%	7.4% (71% of deaths were related to Wilson Disease)	
Alagille syndrome [15]	163	NA	– 9 years (2 months–29 years) – 14 years (3–44 years)	10 years (2 months–44 years)	Of 132 patients with neonatal cholestatic jaundice: 102 were jaundiced, 112 had pruritus, and 40 had xanthomas; 35/76 livers had cirrhosis 25/71 patients had varices Of 31 pts without neonatal cholestatic jaundice: 5 jaundiced, 17 well controlled pruritus, 9 xanthomas; cirrhosis in 6/18, varices in 4/11, no LTx	Survival rates with native liver = 51% and 38% at 10 and 20 y, respectively	LT x in one third patients with neonatal cholestatic jaundice	48 patients with and 9 without cholestatic jaundice died (17 and 2 related to liver disease, respectively)	Overall survival rates were 68% and 62% at 10 and 20 years, respectively
PFIC [16]	61 FIC1 84 BSEP	NA	16–17 years	4.4 years (1.7–9.2), 3.6 years (2.0–8.9)	BSEP pts: gallstones and portal hypertension FIC1 pts: diarrhea, pancreatic disease, rickets, pneumonia, abnormal sweat tests, hearing impairment, poor growth D482G mutation in BSEP pts < rapidly progressive dis	D482G patients survived to a greater age without LTx than did other BSEP or FIC 1 patients	26/61 (43%) FIC1 patients, 38/84 (45%) BSEP patients	7 patients who did not undergo surgical intervention died	The probability of any form of surgery was less in BSEP patients with D482G than in others
Crigler-Najjar [17]	7 (5 type I; 2 type I/II)	18 days–2 months	NA	8.3 years (14 months–17 years)	2 patients developed kernicterus Remaining patients maintained indirect bilirubin from 15 to 25 mg/dL with no associated neurological alterations	NA	2	NA	NA

AIH: Autoimmune hepatitis; BSEP: bile salt export pump; Dx: diagnosis; FU: follow-up; HCC: hepatocellular carcinoma; IBD: inflammatory bowel disease; LTx: liver transplantation; NA: not available; NAFLD: non-alcoholic fatty liver disease; PFIC: progressive familial intra-hepatic cholestasis; plts: platelets; PSC: primary sclerosing cholangitis.

Conclusions

Transition overall is not a simple passage from a paediatric ward to a service dedicated to adults, but is a gradual process for which general rules are still in progress, especially in paediatric hepatology.

Medical problems at the time of transition of young people with chronic hepatobiliary disease are often complex. General healthcare needs and a large number of hepatobiliary disease-specific features may impact on morbidity and mortality in adulthood. Their correct knowledge by patients, adult practitioners and hepatologists responsible for ensuring their continuity of care might help to better manage these conditions they are not usually accustomed to.

Received 8 June 2016; received in revised form 20 November 2016; accepted 21 November 2016

Review

JOURNAL OF HEPATOLOGY

The management of childhood liver diseases in adulthood

Deepak Joshi[1,*], Nitika Gupta[2], Marianne Samyn[1], Maesha Deheragoda[1], Fabienne Dobbels[3], Michael A. Heneghan[1]

Key point

The number of patients with childhood liver diseases surviving into adulthood is increasing. Adult hepatologists need to be familiar with the management of these diseases.

Key point

A sensitive and responsive transition service is required to manage young adults with liver disease.

Key point

Self-management support is an important strategy which empowers young adults to participate in their care.

Key point

Transition services need to be holistic and adopt a multi-disciplinary approach. Successful transition services aim to improve adherence and long-term outcomes

Table 2. Risk factors for non-adherence and approach. Modified from Dobbels *et al.* [20].

	Risk factor	Approach
Socioeconomic factors	Social isolation Family instability Poor parental support Single parent families Cost of medication or clinic visits	Social worker review Review eligibility for financial support.
Patient derived factors	Poor understanding of condition Mental illness Previous non-adherence Past history of child abuse Low self-esteem Post-traumatic stress disorder	Patient passports Clinical psychology review
Disease related factors	Duration of illness Lack of symptoms Substance misuse	Review of clinical status Peer support groups
Treatment related factors	Side effects Number of medications Cost of medication	Regular review of medications
Health care system/health care team factors	Poor communication between the different health care teams, patient and parents Poor relationship between health care teams, patient and parents Lack of continuity of care Clinic attendance resulting in time off school or work	Weekly multi-disciplinary meetings Identification of key care providers Evening clinics

GUT

UK guideline on transition of adolescent and young persons with chronic digestive diseases from paediatric to adult care

Alenka J Brooks, Philip J Smith, Richard Cohen, Paul Collins, Andrew Douds, Valda Forbes, Daniel R Gaya, Brian T Johnston, Patrick J McKiernan, Charles D Murray, Shaji Sebastian, Monica Smith, Lisa Whitley, Lesley Williams, Richard K Russell, Sara A McCartney and James O Lindsay

Gut published online February 21, 2017

SUMMARY OF RECOMMENDATIONS

Recommendations: patient populations involved in adolescent and young person transition

We recommend that adolescent and young person (AYP) with IBD, coeliac disease and chronic liver disease should be involved in formal transition arrangements (Grading of Recommendations Assessment, Development and Evaluation (GRADE) recommendation: strong (C)).

Table 3 The estimated (and extrapolated) UK prevalence of chronic paediatric GI/liver conditions and number of patients transitioned each year using the best available epidemiological, audit and survey data as well as expert opinion

GI/liver condition	UK paediatric prevalence (total population or proportion within populations)	Estimated number of patients transitioned per year
IBD*	7000	1000 (~60% Crohn's disease)
Chronic liver disease	The prevalence of paediatric liver disease in England between 2008 and 2014: 11–15 years: 1502 16–17: 624†[17]	Approximately 61–76 transplant cases per annum‡[18] 753 paediatric patients were transferred to adult services between 2008 and 2014) (proportion which are non-transplant is unknown)†[17]
Complex enteral	No relevant data identified	No relevant data identified
Parenteral nutrition (combination of long term in and out of hospital)	290§	60
Coeliac disease	3–13 per 1000 children, or approximately 1:80 to 1:300 children	600 per year
Allergic/eosinophilic oesophagitis	Food allergy 0.6%[4] Eosinophilic oesophagitis 0.2 per 100 000	No specific data
Functional GI disease	~10%[5]	No specific data

*Estimates derived from the organisational IBD audit 2013 and local figures (RKR).
†Williams *et al.*[17]
‡Data from annual report on liver transplantation NHS England September 2014 (2004–2014).
§Data courtesy of Henry Gowens/Andy Barclay British Intestinal Failure Survey data (unpublished 2014).

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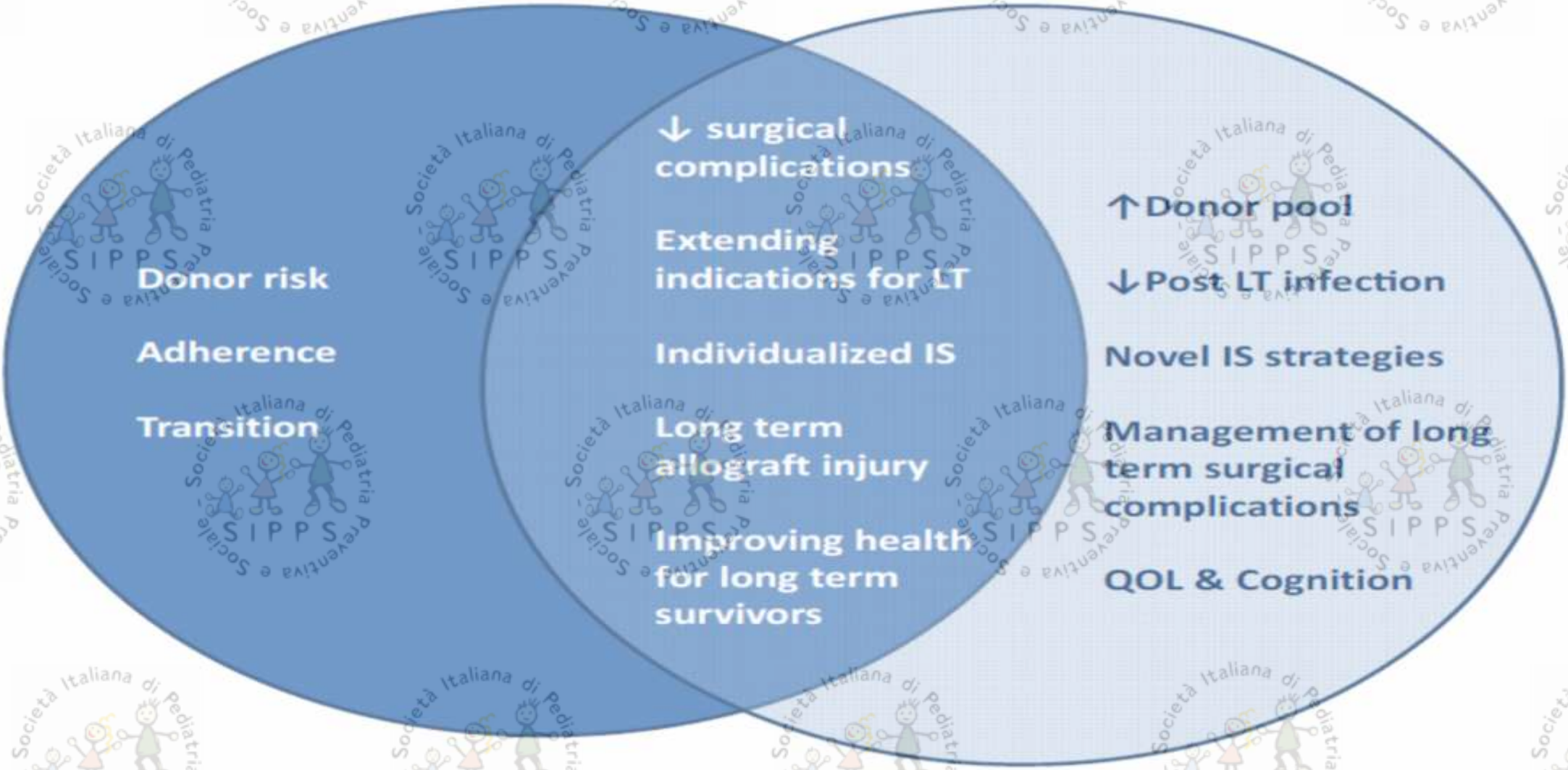
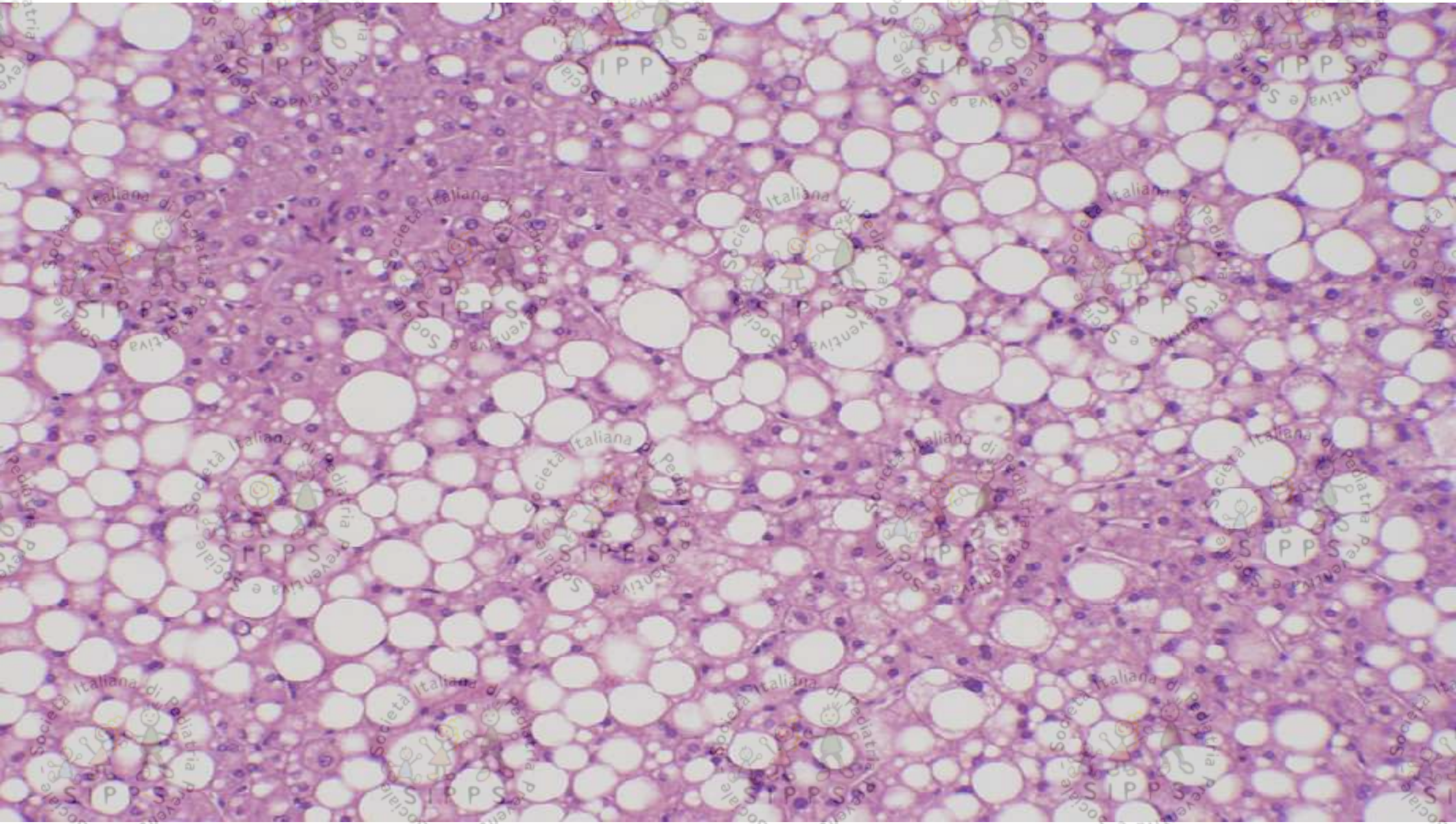
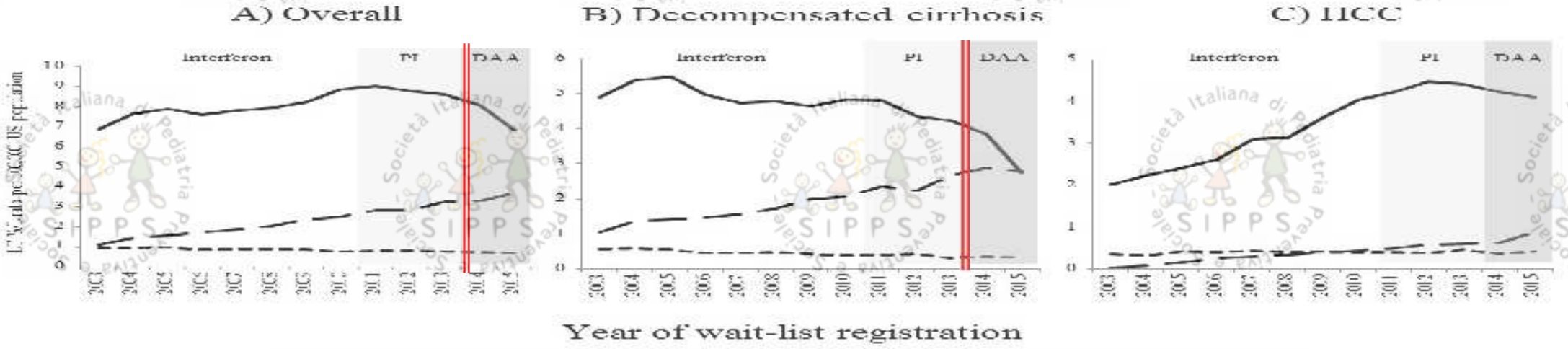
Early and late factors impacting patient and graft outcome in pediatric liver transplantation: summary of an ESPGHAN Monothematic Conference

Valérie A. McLin[1]*, Upton Allen[2], Olivia Boyer[3], John Bucuvalas[4], Michele Colledan[5], Maria-Cristina Cuturi[6], Lorenzo d'Antiga[7], Dominique Debray[8], Antal Dezsofi[9], Jean de Ville de Goyet[10], Anil Dhawan[11], Ozlem Durmaz[12], Christine Falk[13], Sandy Feng[14], Björn Fischler[15], Stéphanie Franchi-Abella[16], Esteban Frauca[17], Rainer Ganschow[18], Stephen Gottschalk[19], Nedim Hadzic[20], Loreto Hierro[17], Simon Horslen[21], Stefan Hubscher[22], Vincent Karam[23], Deirdre Kelly[24], Britta Maecker-Kolhoff[25], George Mazariegos[26], Patrick McKiernan[24], Anette Melk[27], Valerio Nobili[28], Funda Ozgenç[29], Raymond Reding[30], Marco Sciveres[31], Khalid Sharif[24], Piotr Socha[32], Christian Toso[33], Pietro Vajro[34], Anita Verma[35], Barbara E. Wildhaber[1], Ulrich Baumann[36]


Donor risk
Adherence
Transition
↓ surgical complications
Extending indications for LT
Individualized IS
Long term allograft injury
Improving health for long term survivors
↑Donor pool
↓Post LT infection
Novel IS strategies
Management of long term surgical complications
QOL & Cognition
SPLIT
ESPGHAN





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OCCHI SUL DIABETE

TIENI GLI OCCHI APERTI SU

In pratica oggi il 9,2% della popolazione italiana ha difficoltà a mantenere sotto controllo la glicemia.

Nel 2030 si prevede che le persone diagnosticate con diabete tipo 2 saranno 5 milioni.

popolazione
1 milione di persone abbiano il diabete di tipo 2 ma non siano state diagnosticate: è l'1,6% della popolazione
2,6 milioni di persone abbiano difficoltà a mantenere le glicemie nella norma, una condizione che nella maggior parte dei casi prelude allo sviluppo del diabete di tipo 2. Parliamo del 4,3% della popolazione

Scopri la campagna di informazione sul diabete realizzata per l'edizione 2016 della Giornata Mondiale del Diabete.

Campione: 20105457
Ingresso: 23/12/14 08:05.20
Data di Nascita: 15/11/2004
ID Paziente:
Dottore:
Diagnosi:

Nom
Cogn
Repa
C.F.:
Data
Ora

CHIMICA CLINICA

Resp.: Prof.s. orzio

UOC BIOCHIMICA CL
Din

CURVA GLICEMICA DA CARICO

GLICEMIA		
Glicemia +30 min	81	mg/d
Glicemia +60 min	138	mg/d
Glicemia +90 min	163	mg/d
Glicemia +120 min	161	mg/d
	129	mg/d

Refertato e Firmato da: Dott.ssa Annamaria De Angelis

ENDOCRINOLOGIA

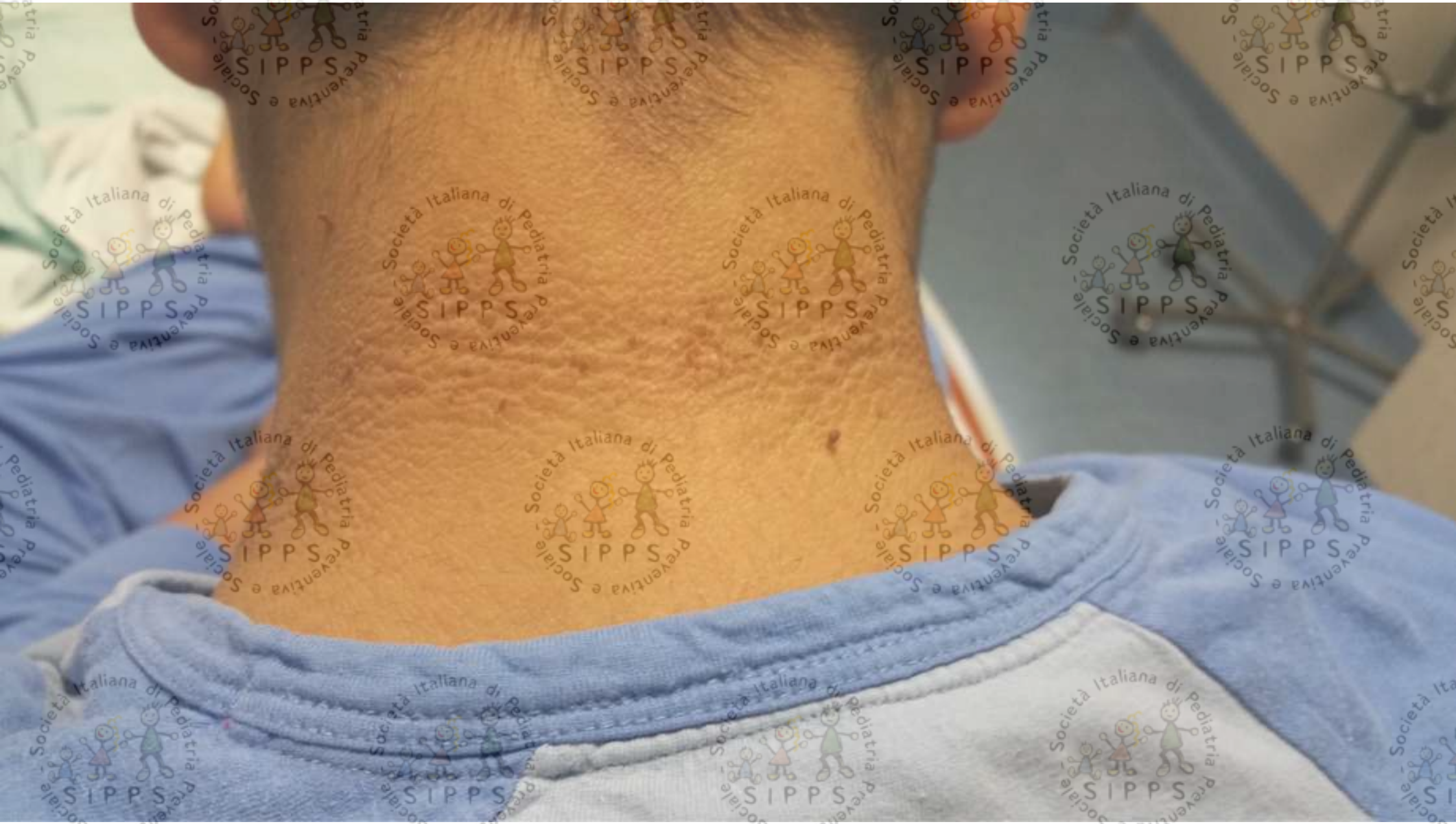
Resp.: Prof.ssa Ottavia Porzio

INSULINA		
Insulina 30 min	43.07	µU/m
Insulina 60 min	215.74	µU/m
Insulina 90 min	383.12	µU/m
Insulina 120 min	450.98	µU/m
	295.77	µU/m

Refertato e Firmato da:

Roma, mercoledì 31 dicembre 2014

Il R



Bambino Gesù
OSPEDALE PEDIATRICO

Ospedale Pediatrico Bambino Gesù

ISTITUTO DI RICOVERO E CURA A CARATTERE SCIENTIFICO

Laboratorio Analisi - Sede di Roma tel. 06/68591

Primario Prof. Maurizio Muraca

Joint Commission International

Data di Stampa: 19/12/2014 Ore: 14:50 Pag.: 1

Id.:02403821 Sig.

Nosologico: **2014150780**

Data Nascita: **28/02/1998** Età: 16 Anni

Sesso M

Provenienza: 000004671010 RM Malattie Epato-Metaboliche RO

Routine

Richiesta: 12193977 **19/12/2014** Ore: 08:00

Esame	*Esito*	*U.M.*	*Valori Riferimento*
ENDOCRINOLOGIA2			
Insulina Basale (met. Chemilum.)	**29.10**	mU/L	3.00 - 25.00
Insulina punto 4	**198.2**	mU/L	
CHIMICA CLINICA DI BASE			
CARICO/STIMOLO GLICEMICO			
Glicemia Basale	**86**	mg/dl	55 - 110
Glicemia +120	**146**	mg/dl	

Bambino Gesù
OSPEDALE PEDIATRICO

Ospedale Pediatrico Bambino Gesù

ISTITUTO DI RICOVERO E CURA A CARATTERE SCIENTIFICO
Laboratorio Analisi - Sede di Roma tel. 06/68591
Primario Prof.ssa Ottavia Porzio

Joint Commission International

Data di Stampa: 06/11/2015 Ore: 11:01 Pag.: 1

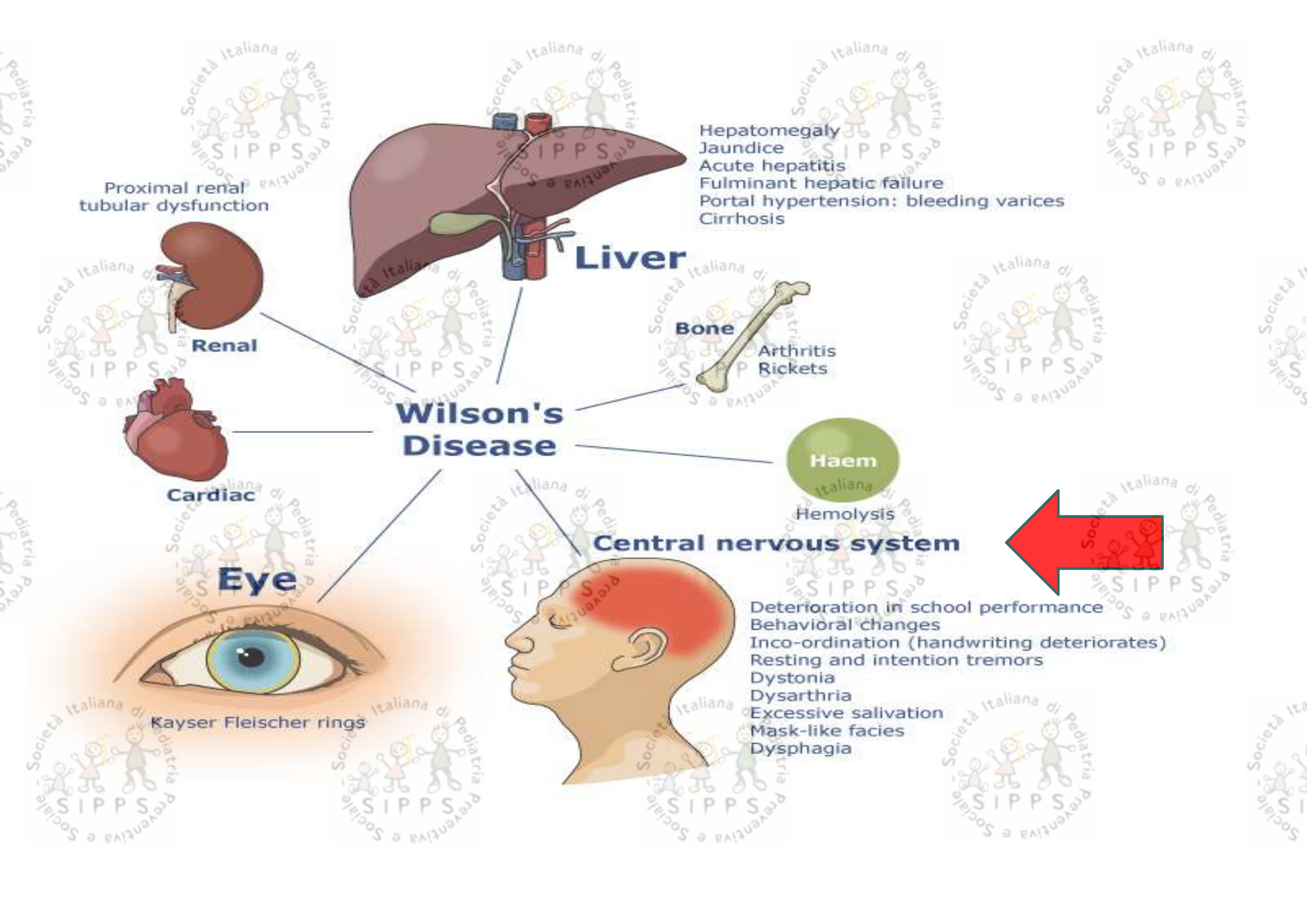
Id: 02403821 Sig.
Nosologico: 2015151105
Data Nascita: 28/02/1998 Età: 17 Anni
Sesso: M

Provenienza: 000004671010 RM Malattie Epato-Metaboliche RO

Routine

Richiesta: 11053473 05/11/2015 Ore: 08:50

Esame	*Esito*	*U.M.*	*Valori Riferimento*
ENDOCRINOLOGIA2			
Insulina Basale (met. Chemilum.)	34.33 >	mU/L	3.00 - 25.00
Insulina punto 4	556.3	mU/L	
Valore ottenuto con diluizione del campione			
CHIMICA CLINICA DI BASE			
CARICO/STIMOLO GLICEMICO			
Glicemia Basale	87	mg/dL	55 - 110
Glicemia +120	212	mg/dL	


Hepatomegaly
Jaundice
Acute hepatitis
Fulminant hepatic failure
Portal hypertension: bleeding varices
Cirrhosis
Liver
Proximal renal tubular dysfunction
Renal
Bone
Arthritis
Rickets
Wilson's Disease
Cardiac
Haem
Hemolysis
Central nervous system
Eye
Kayser Fleischer rings
Deterioration in school performance
Behavioral changes
Inco-ordination (handwriting deteriorates)
Resting and intention tremors
Dystonia
Dysarthria
Excessive salivation
Mask-like facies
Dysphagia

Parkinsonism and Related Disorders 24 (2016) 15–19

Contents lists available at ScienceDirect

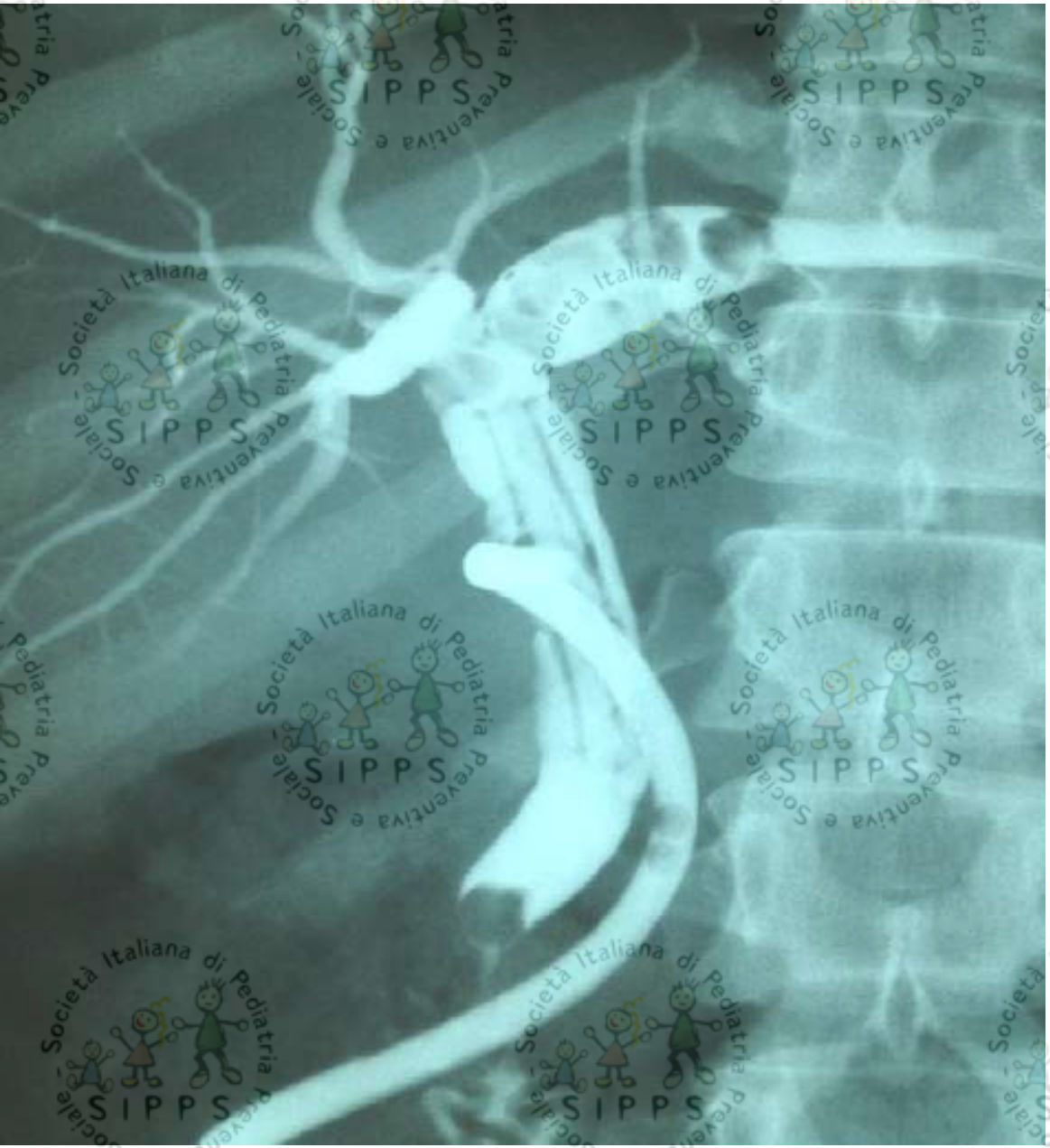
Parkinsonism and Related Disorders

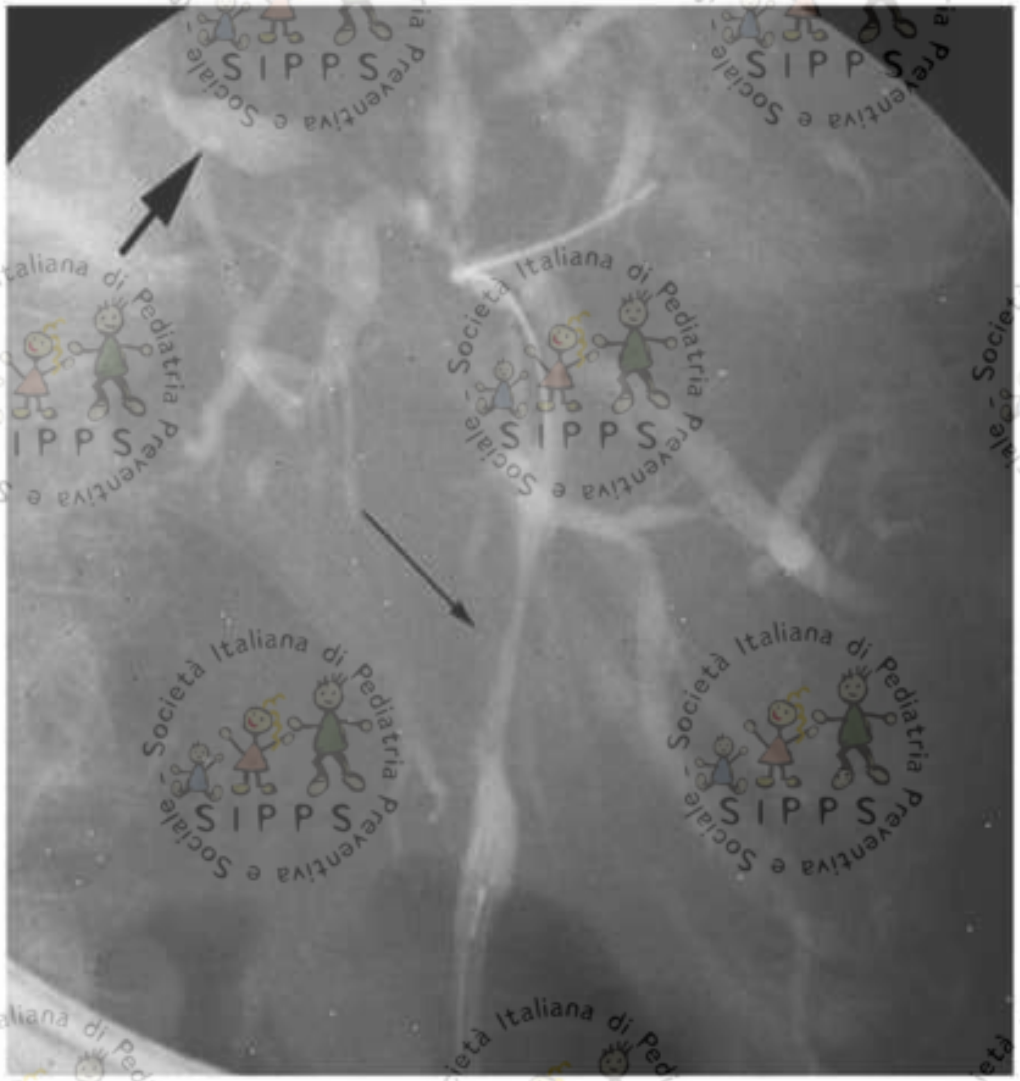
journal homepage: www.elsevier.com/locate/parkreldis

Subclinical neurological involvement does not develop if Wilson's disease is treated early

Raffaele Dubbioso [a, 1], Giusy Ranucci [b, 1], Marcello Esposito [a, 1], Fabiola Di Dato [b], Antonietta Topa [a], Mario Quarantelli [c], Margherita Matarazzo [d], Lucio Santoro [a], Fiore Manganelli [a, 2], Raffaele Iorio [b, *, 2]

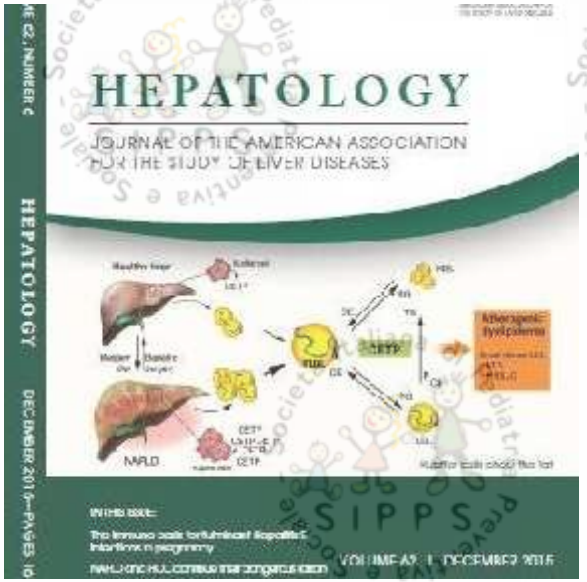
[a] Department of Neurosciences, Reproductive and Odontostomatological Sciences, University of Naples Federico II, Italy
[b] Department of Translational Medical Sciences, Section of Pediatrics, University of Naples Federico II, Italy
[c] Institute of Biostructure and Bioimaging, National Research Council (CNR), Naples, Italy
[d] Department of Translational Medical Sciences, Section of Internal Medicine, University of Naples Federico II, Italy





Visione alla CPRE di tratti biliari stenotici e tratti dilatati in Colangite Sclerosante.

The natural history of primary sclerosing cholangitis in 781 children: a multicenter, international collaboration



Corresponding Author:

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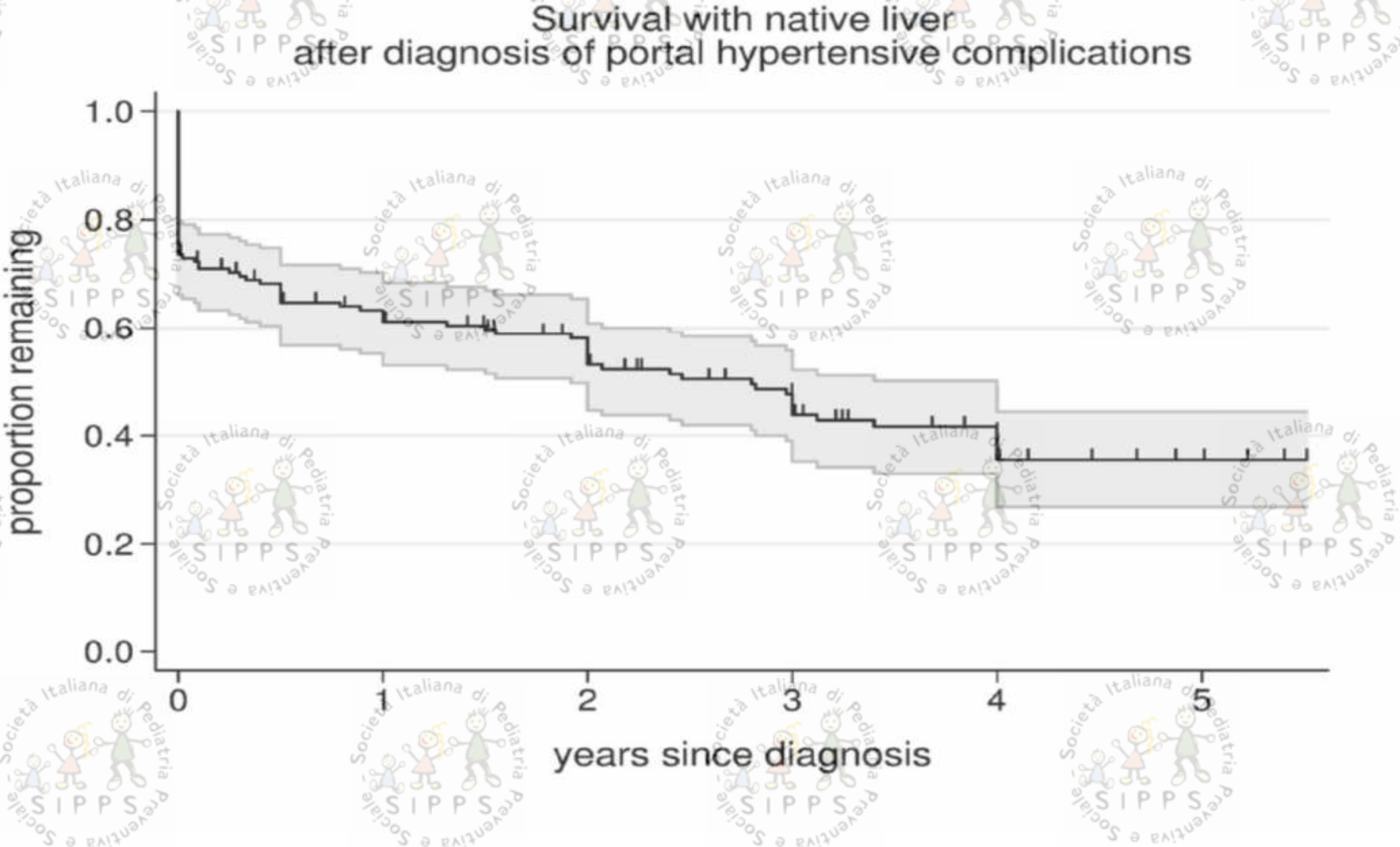
(Fax) 801-587-7539


Survival with native liver
after diagnosis of portal hypertensive complications
proportion remaining
1.0
0.8
0.6
0.4
0.2
0.0
0
1
2
3
4
5
years since diagnosis

26th May 10:10–10:25	Prof. John J. Gargus Human Genetics, Pediatrics, Physiology & Bio UCI Irvine, Irvine, California, USA
26th May 16:20–16:35	Prof. Valerio Nobili Head of Hepato-Metabolic Liver Disease secti and Chief of the Liver Research Unit, Bambino Gesù Children's Hospital, Rome, Ital
27th May 10:50–11:05	Prof. John J. Gargus
27th May 13:45–14:00	Prof. Valerio Nobili

WHAT IS YOUR LIKELY DIAGNOSIS?

Challenging Presentation in Paediatric Hepatic Disease

Advancing Treatment in Lysosomal Acid Lipase Deficiency:

James Underberg, MD, MS; Don P. Wilson, MD; Patrick M. Moriarty, MD Faculty and Disclosures

CME/CE Released: 6/27/2015; Valid for credit through 6/27/2016

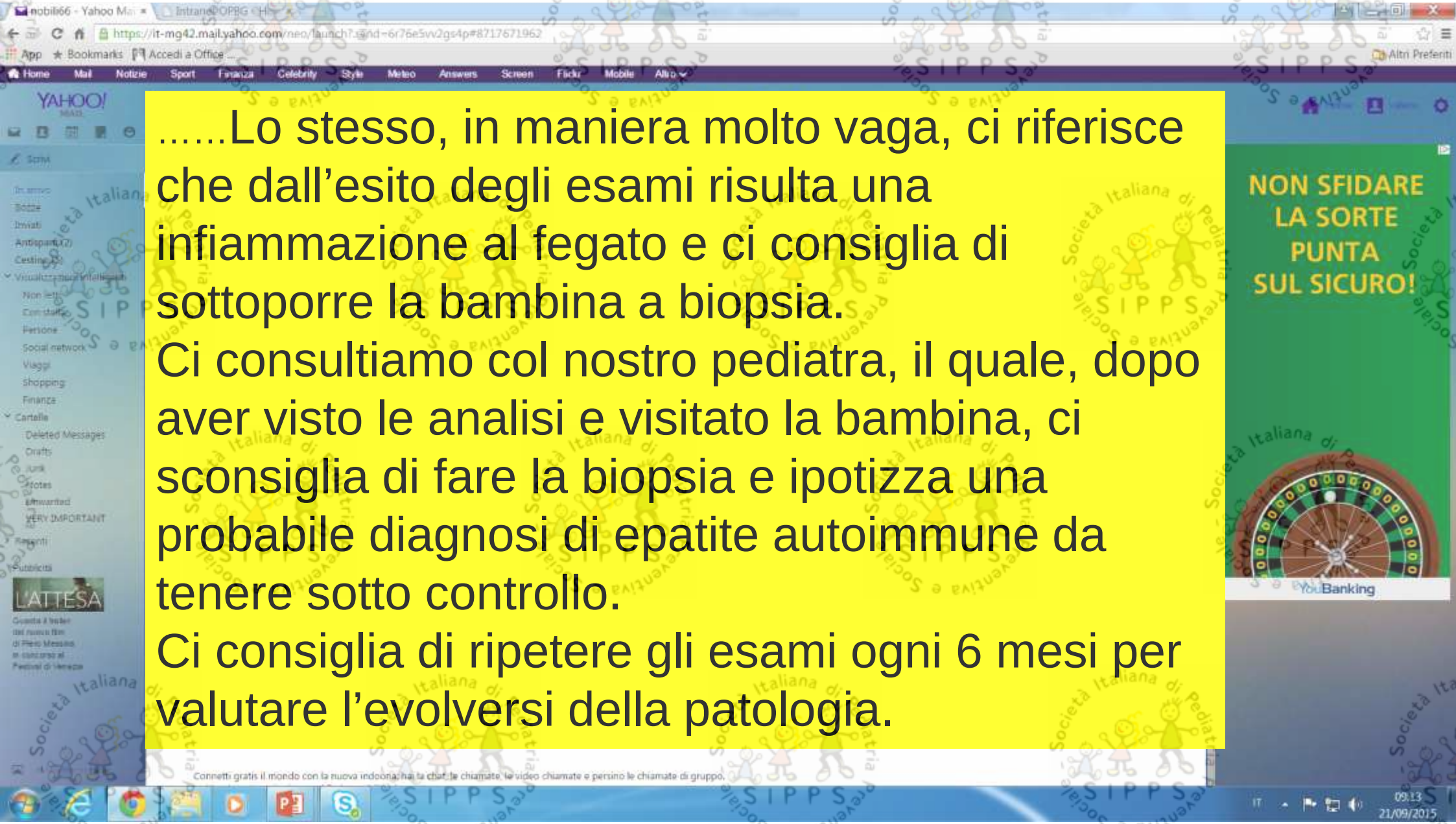
Slides/Video

How confident are you in diagnosing lysosomal acid lipase deficiency (LAL-D)?

	Your Colleagues Responded:
Very confident	10%
Somewhat confident	22%
Not confident	78%

How confident are you in treating LAL-D?

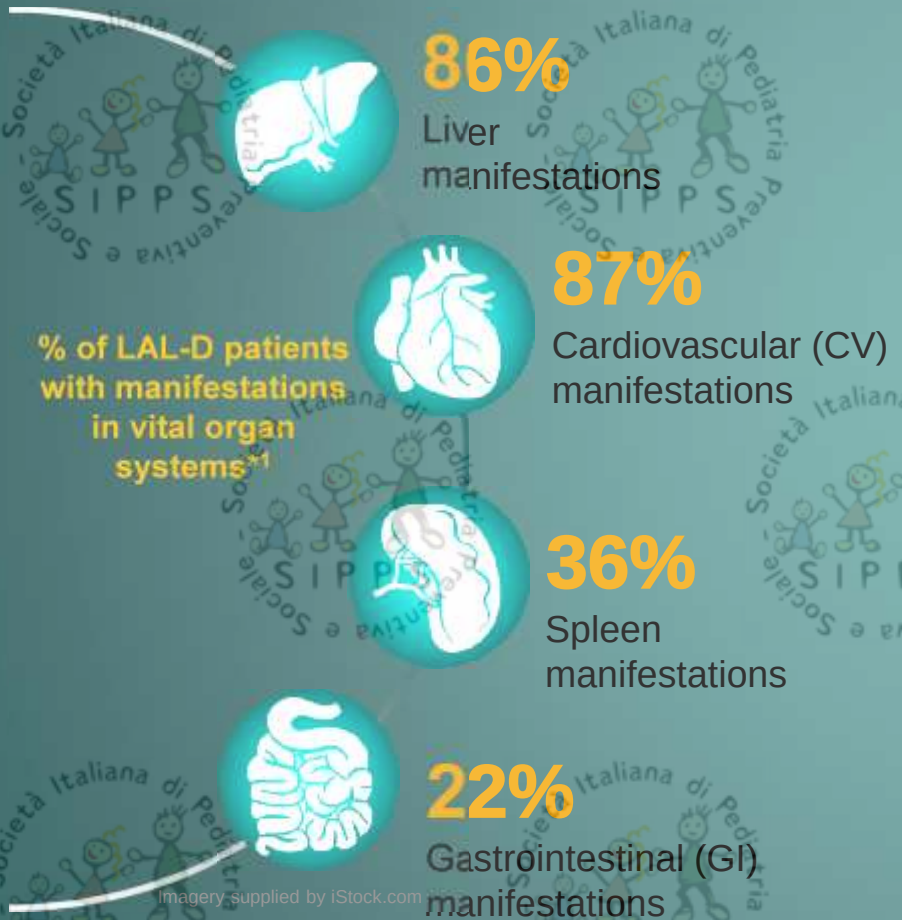
	Your Colleagues Responded:
Very confident	10%
Somewhat confident	21%
Not confident	78%


.....Lo stesso, in maniera molto vaga, ci riferisce che dall'esito degli esami risulta una infiammazione al fegato e ci consiglia di sottoporre la bambina a biopsia.
Ci consultiamo col nostro pediatra, il quale, dopo aver visto le analisi e visitato la bambina, ci sconsiglia di fare la biopsia e ipotizza una probabile diagnosi di epatite autoimmune da tenere sotto controllo.
Ci consiglia di ripetere gli esami ogni 6 mesi per valutare l'evolversi della patologia.
NON SFIDARE LA SORTE PUNTA SUL SICURO!
YouBanking

Example 2: Early Onset Lysosomal Acid Lipase Deficiency (Wolman)

- Prominent hepatic and GI manifestations
  - Hepatomegaly and liver failure
  - Splenomegaly
  - Persistent vomiting
  - Abdominal distension
  - Profound growth failure
- Adrenal calcification
- Rapidly progressive and fatal

LAL-D is a life-threatening genetic disease associated with significant morbidities and increased risk for premature mortality[1,2]



LAL-D in children and adults[1,2]

- Progressive disease due to lysosomal accumulation of CE and TG. Results in liver disease and dyslipidaemia
- Clinical manifestations
  - Fibrosis and cirrhosis
  - Liver failure requiring transplantation
  - Dyslipidaemia leading to accelerated atherosclerosis
- 87% of patients showed manifestations in more than 1 organ system

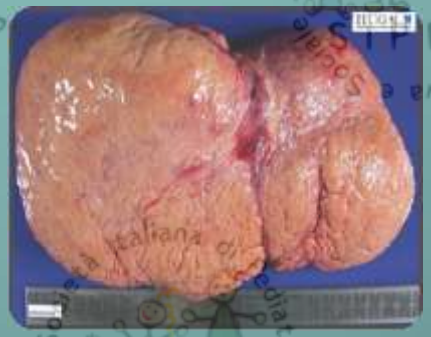


Image © Courtesy of Gazi University Hospital

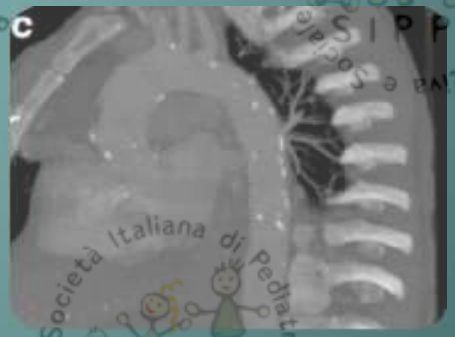


Image from Ambler GK, *et al. JIMD Rep* 2013;8:41–6.

*Based on an analysis of 55 genotyped LAL-D patients in a cohort of 135 cases

1. Bernstein DL, *et al. J Hepatol*. 2013;58(6):1230–43; 2. Reiner Ž, *et al. Atherosclerosis*. 2014;235(1):21–30.

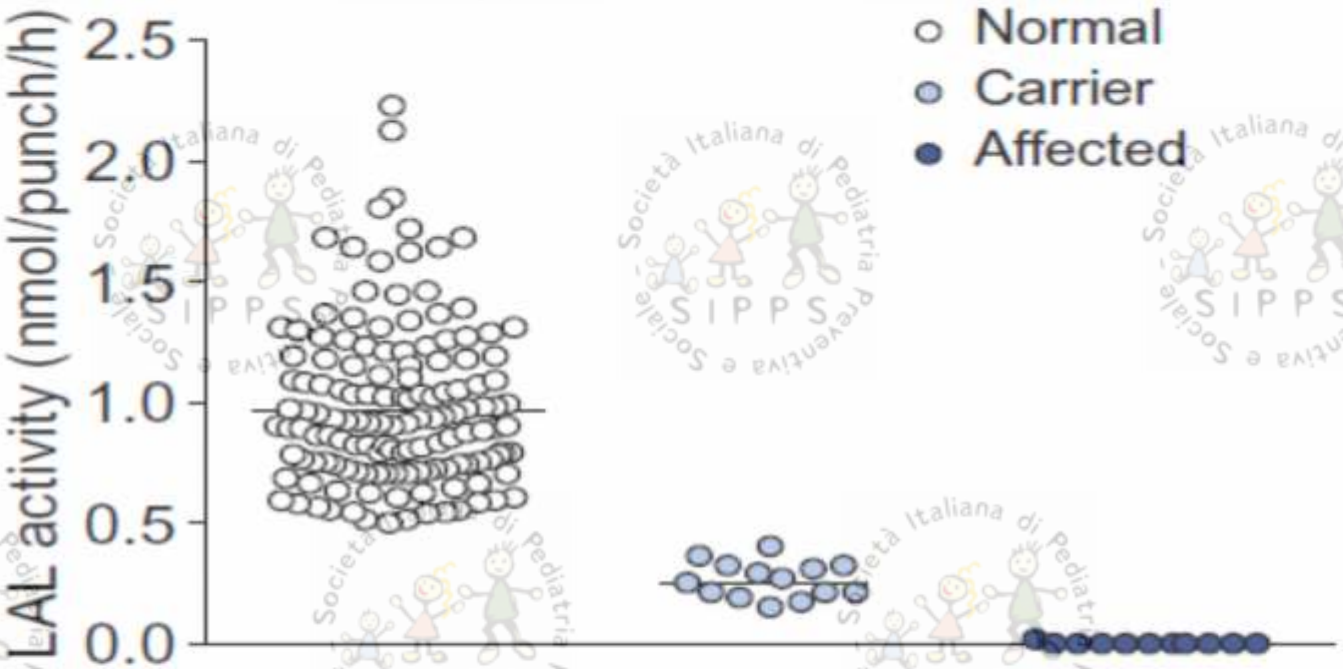
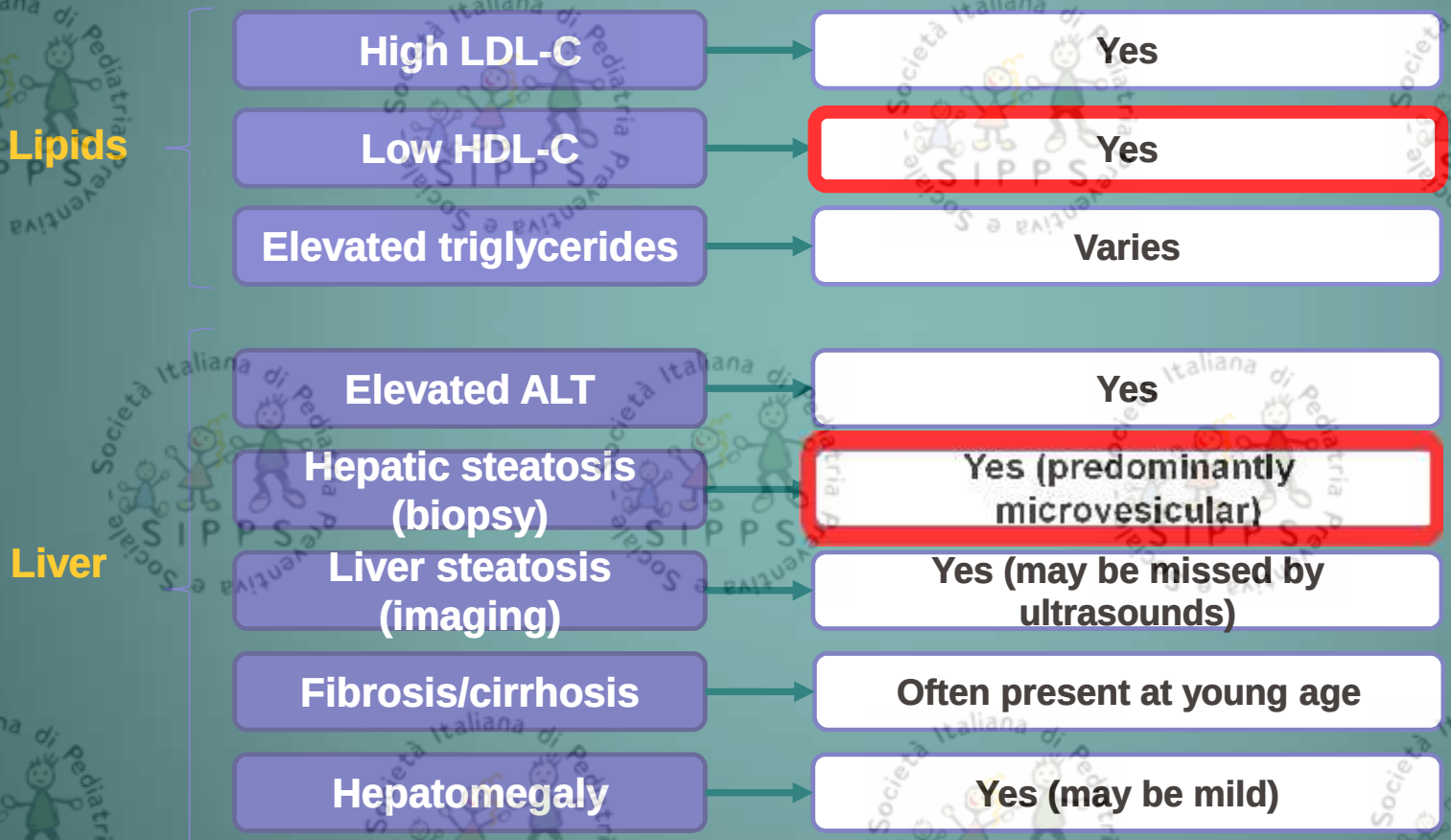


Fig. 3. Dried blood spot LAL activity in affected homozygotes, heterozygotes, and normal individuals. This assay is LAL specific, as it incorporates a specific inhibitor, Lalistat 2, which reduces the activity of the other lipases. LAL activity in 140 normal controls, 11 LAL deficient patients and 15 carriers. Limit of detection = 0.02 nmol/punch/h. From reference [109], with permission.

LAL-D: differential diagnosis



1. Reiner Ž, et al. *Atherosclerosis.* 2014;235(1):21–30. 2. Bernstein DL, et al. *J Hepatol.* 2013;58:1230–43. doi:10.1016/j.jhep.2013.02.014.

LAL-D: differential diagnosis

	Similarity to LAL-D	Consider LAL-D
Wilson's disease	Asymptomatic hepatomegaly, isolated splenomegaly, persistently elevated AST, ALT, fatty liver, cirrhosis[1]	Patient does not have signs of CNS involvement[2]
Metabolic syndrome	Patients have decreased HDL-c and elevated TG[3]	Patient has signs of metabolic syndrome and dyslipidaemia[3]
FCH	Patients have decreased HDL-c and elevated LDL-c[3]	Patient presents with lab values similar to FCH with ALT > ULN[3]
HeFH	Patients have elevated LDL-c[3,4]	Patient presents with lab values similar to HeFH, with ALT > ULN, or no confirmed mutation for HeFH-related gene[3,4]
NAFLD	Some patients may have elevated ALT, with or without hepatic steatosis[5]	Patient has NAFLD/NASH with ALT > ULN, **and** dyslipidaemia[3]

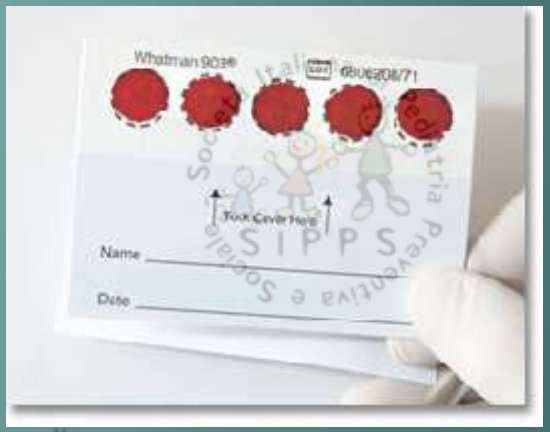
FCH, familial combined hyperlipidaemia; HeFH, heterozygous familial hypercholesterolaemia

1. Roberts EA and Schilsky ML. *AASLD Practice Guidelines. Hepatology*. 2008;47(6):2089. 2. Manolaki N, *et al. J Pediatr Gastroenterol Nutr* 2009;48(1):72–7. 3. Reiner Ž, *et al. Atherosclerosis* 2014;235(1):21–30; 4. Varghese M. *Ann Pediatr Cardiol* 2014;7(2):107–17; 5. Bhala N, *et al. Curr Pharm Des*

In all patients:

- Laboratory assay
- Screening for Metabolic Syndrome
- Lysosomal Acid Lipase assay

Blood was spotted on to filter paper on the day of venipuncture and allowed to dry overnight at room temperature.



- 168 patients underwent to Liver biopsy

A reference range was established using blood submitted to our laboratory for routine diagnostic testing (0,73 nmol).

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

A Phase 3 Trial of Sebelipase Alfa in Lysosomal Acid Lipase Deficiency

E.K. Burton, M. Balwani, F. Feillet, I. Barić, T.A. Burrow, C. Camarena Grande, M. Coker, A. Consuelo-Sánchez, P. Deegan, M. Di Rocco, G.M. Enns, R. Erbe, F. Ezgu, C. Ficicioglu, K.N. Ezuruya, J. Kane, C. Laukaitis, E. Mengel, E.G. Neilan, S. Nightingale, H. Peters, M. Scarpa, K.O. Schwab, V. Smolka, V. Valayannopoulos, M. Wood, Z. Goodman, Y. Yang, S. Eckert, S. Rojas-Caro, and A.G. Quinn

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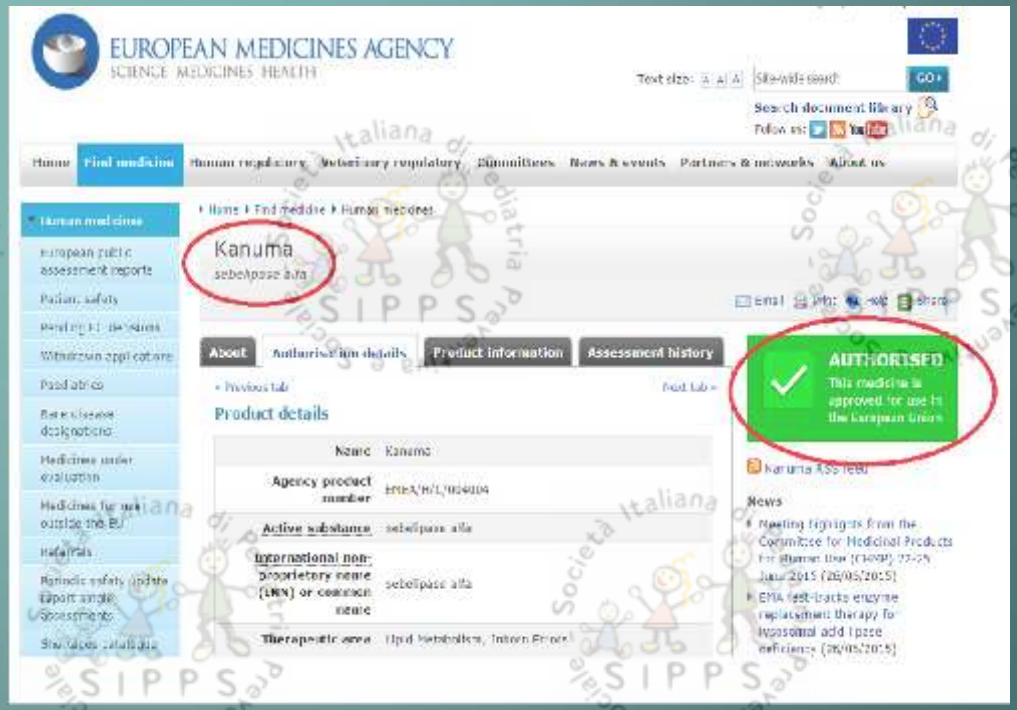
FDA News Release

FDA approves first drug to treat a rare enzyme disorder in pediatric and adult patients

SHARE | TWEET | LINKEDIN | PIN IT | EMAIL | PRINT

For Immediate Release: December 8, 2015

Release: Today, the U.S. Food and Drug Administration approved Kanuma (sebelipase alfa)



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Kanuma
sebelipase alfa

About | Authorisation details | Product information | Assessment history

AUTHORISED
This medicine is approved for use in the European Union

Product details

Name	Kanuma
Agency product number	EMEA/H/C/004004
Active substance	sebelipase alfa
International non-proprietary name (INN) or common name	sebelipase alfa
Therapeutic area	Lipid Metabolism, Inborn Errors

News

- Meeting highlights from the Committee for Medicinal Products for Human Use (CHMP) 22-25 June 2015 (26/06/2015)
- EMA fast-tracks enzyme replacement therapy for lysosomal acid lipase deficiency (26/06/2015)

Digestive and Liver Disease 48 (2016) 909–913

Contents lists available at ScienceDirect

Digestive and Liver Disease

journal homepage: www.elsevier.com/locate/dld

Liver, Pancreas and Biliary Tract

Reduced lysosomal acid lipase activity – A potential role in the pathogenesis of non alcoholic fatty liver disease in pediatric patients

Praveen Kumar Conjeevaram Selvakumar [a], Mohammad Nasser Kabbany [a], Rocio Lopez [b], Giulia Tozzi [d], Anna Alisi [c], Naim Alkhouri [a,b,*], Valerio Nobili [c,**]

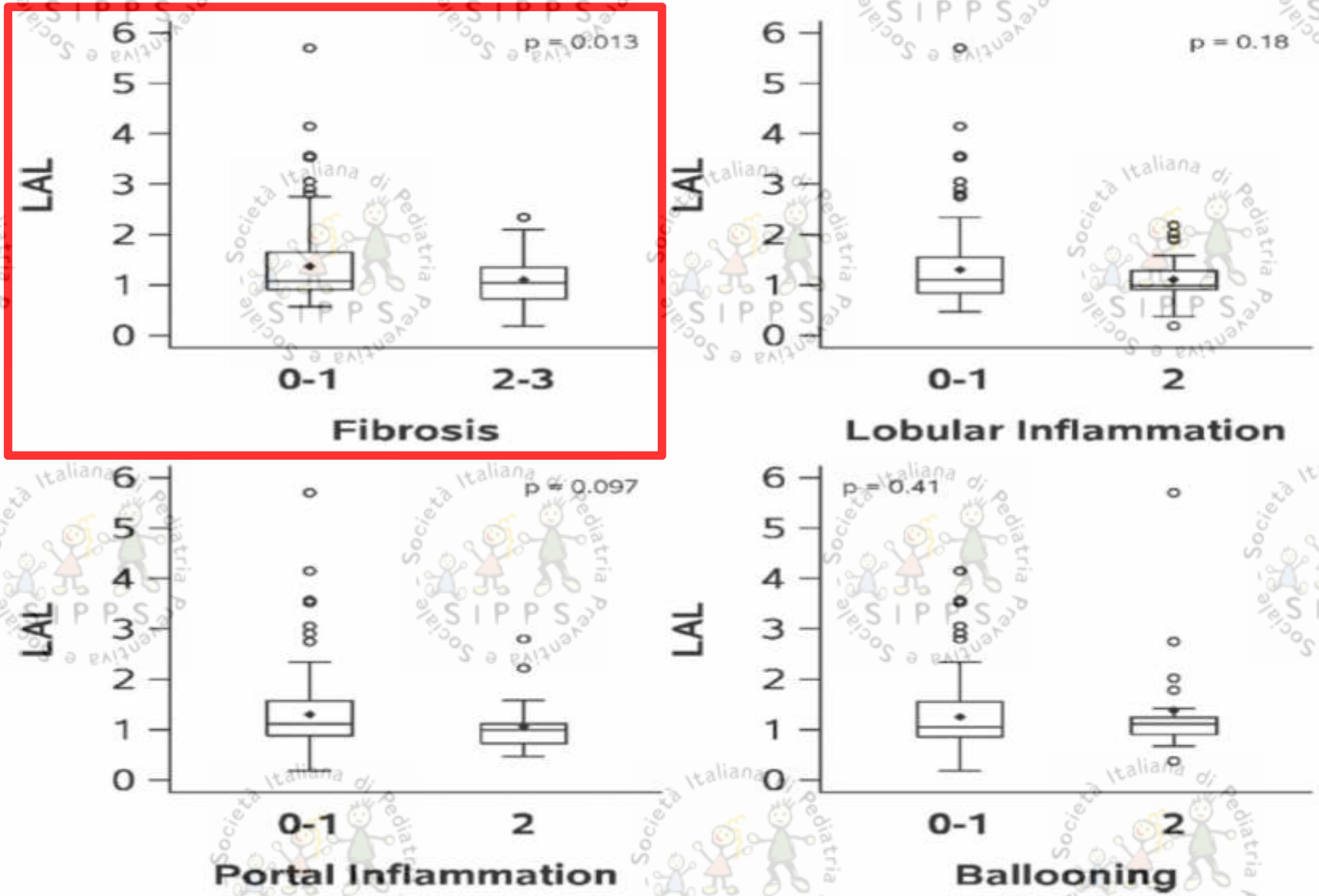

Fig. 1. Box and whisker plot showing association between lysosomal acid lipase activity and individual histology features. LAL, lysosomal acid lipase activity.

Table 3
Associations between lysosomal acid lipase activity and histological features.

Feature Presence	NASH[a]	Fibrosis stages 2–3	Steatosis grades 2–3	Lobular inflammation grades 1–2	Portal inflammation grade 2	Ballooning grade 2
No	1.3 ± 0.57	1.4 ± 0.80	1.2 ± 0.54	1.3 ± 0.73	1.3 ± 0.72	1.2 ± 0.63
Yes	1.2 ± 0.80	1.10 ± 0.45	1.3 ± 0.75	1.1 ± 0.47	1.05 ± 0.51	1.4 ± 1.08
p-Value	0.57	0.013	0.85	0.18	0.097	0.41

Values presented as mean ± SD.
p-Values correspond to Student's *t*-tests.
[a] Nonalcoholic steatohepatitis.

Table 4
Associations between lysosomal acid lipase activity and histological features after adjusting for age, body mass index, triglycerides and cholesterol.

Feature presence	NASH [a]	Fibrosis stages 2–3	Steatosis grades 2–3	Lobular inflammation grades 1–2	Portal inflammation grade 2	Ballooning grade 2
No	1.3 (1.1, 1.4)	1.4 (1.2, 1.5)	1.2 (1.02, 1.4)	1.3 (1.2, 1.4)	1.3 (1.2, 1.4)	1.2 (1.1, 1.4)
Yes	1.3 (1.1, 1.4)	1.10 (0.93, 1.3)	1.3 (1.2, 1.4)	1.07 (0.82, 1.3)	1.07 (0.80, 1.3)	1.4 (1.09, 1.7)
p-Value	0.79	0.015	0.65	0.097	0.13	0.42

Values presented as mean (95% CI).
p-Values correspond to ANCOVA.
[a] Nonalcoholic steatohepatitis.

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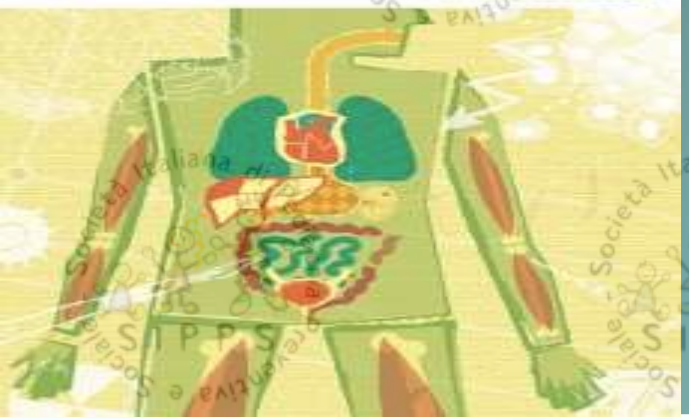


2 | 2015 | VOLUME 1

PRIMER

Nonalcoholic fatty liver disease

Elizabeth M. Brunt[1], Vincent W.-S. Wong[2], Valerio Nobili[3], Christopher P. Day[4], Silvia Sookoian[5], Jacquelyn J. Maher[6], Elisabetta Bugianesi[7], Claude B. Sirlin[8], Brent A. Neuschwander-Tetri[9] and Mary E. Rinella[10]

Published online 17 December 2015

Box 1 | **Steatosis and elevated liver tests**

Drug use

- Tamoxifen
- Amiodarone
- Glucocorticoids
- Synthetic oestrogens
- Antiviral agents (for example, highly active antiretroviral therapies)
- Methotrexate

Other metabolic or genetic causes

- Hypobetalipoproteinaemia
- Lysosomal acid lipase partial deficiency
- Lipodystrophy
- Weber–Christian disease

Nutrition

- Malnutrition
- Malabsorption
- Total parenteral nutrition
- Rapid weight loss
- Jejunoileal bypass

Others

- Small bowel diverticulosis
- Exposure to petrochemicals
- Exposure to organic solvents

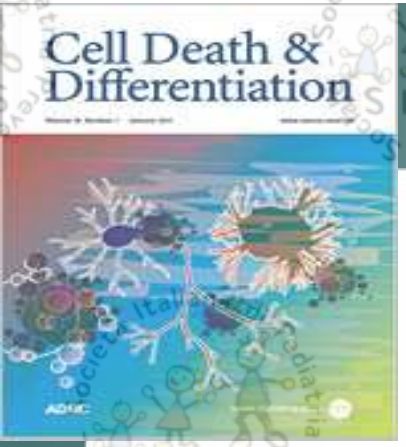
PEDIATRICS
OFFICIAL JOURNAL OF THE AMERICAN ACADEMY OF PEDIATRICS

Pediatrics. 2016 Oct;138(4). pii: e20160214. Epub 2016 Sep 13

Lysosomal Acid Lipase Deficiency Unmasked in Two Children With Nonalcoholic Fatty Liver Disease

Ryan W. Himes, MD,[a] Sarah E. Barlow, MD, MPH,[a] Kevin Bove, MD,[b] Norma M. Quintanilla, MD,[c] Rachel Sheridan, MD,[b] Rohit Kohli, MBBS, MS[d]


SMS
SMS

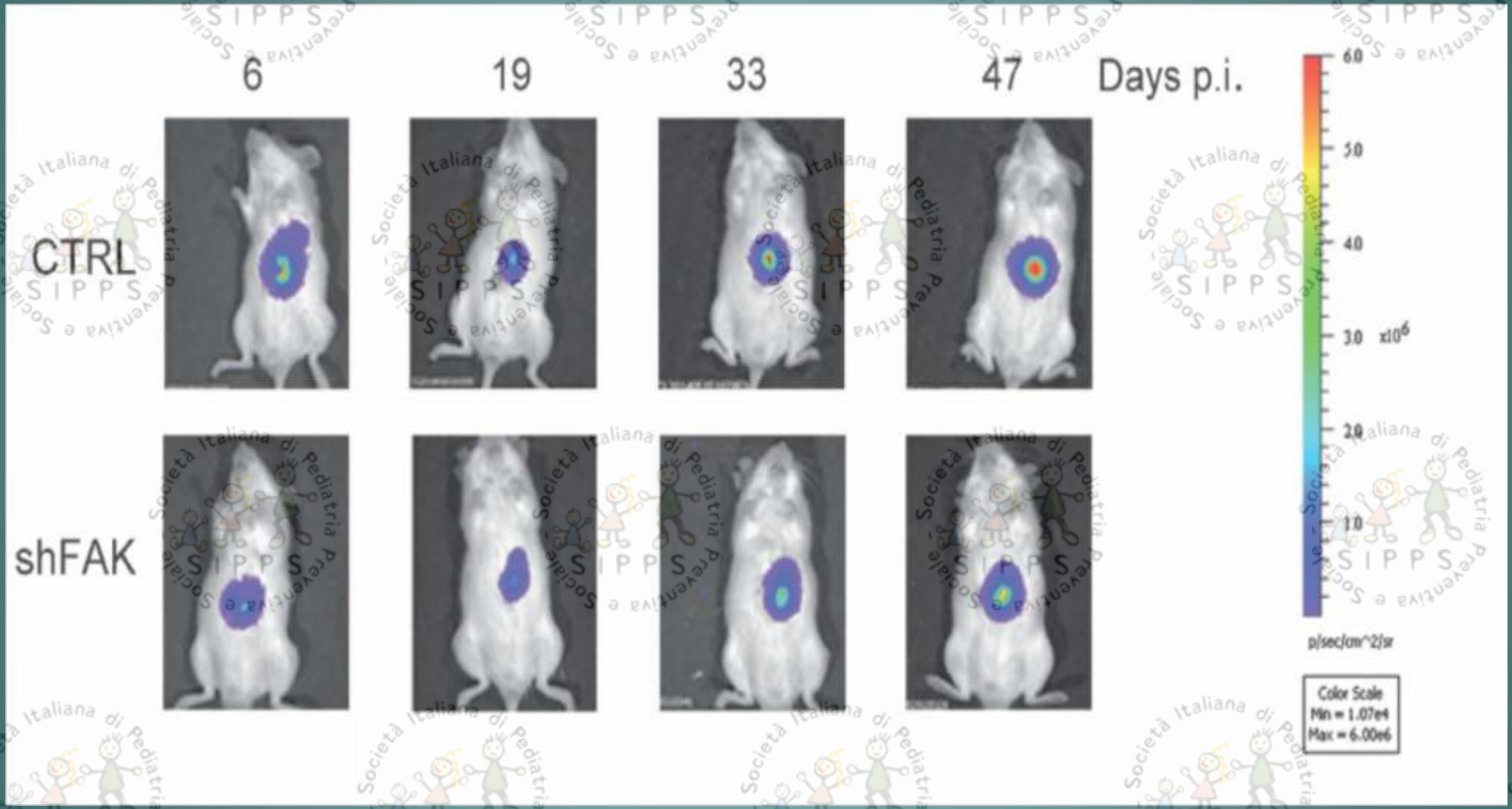


Cell Death and Differentiation (2017), 1–14
Official journal of the Cell Death Differentiation Association

www.nature.com/cdd

Focal adhesion kinase depletion reduces human hepatocellular carcinoma growth by repressing enhancer of zeste homolog 2

Daniela Gnani[1], Ilaria Romito[1], Simona Artuso[2], Marco Chierici[3], Cristiano De Stefanis[1], Nadia Panera[1], Annalisa Crudele[1], Sara Ceccarelli[1], Elena Carcarino[4], Valentina D'Oria[5], Manuela Porru[2], Ezio Giorda[6], Karin Ferrari[7], Luca Miele[8], Erica Villa[9], Clara Balsano[10], Diego Pasini[7], Cesare Furlanello[3], Franco Locatelli[4,11], Valerio Nobili[12], Rossella Rota[4], Carlo Leonetti[2] and Anna Alisi*[,1]


6
19
33
47
Days p.i.
CTRL
shFAK
6.0
5.0
4.0
3.0 x10^6
2.0
1.0
p/sec/cm^2/sr
Color Scale
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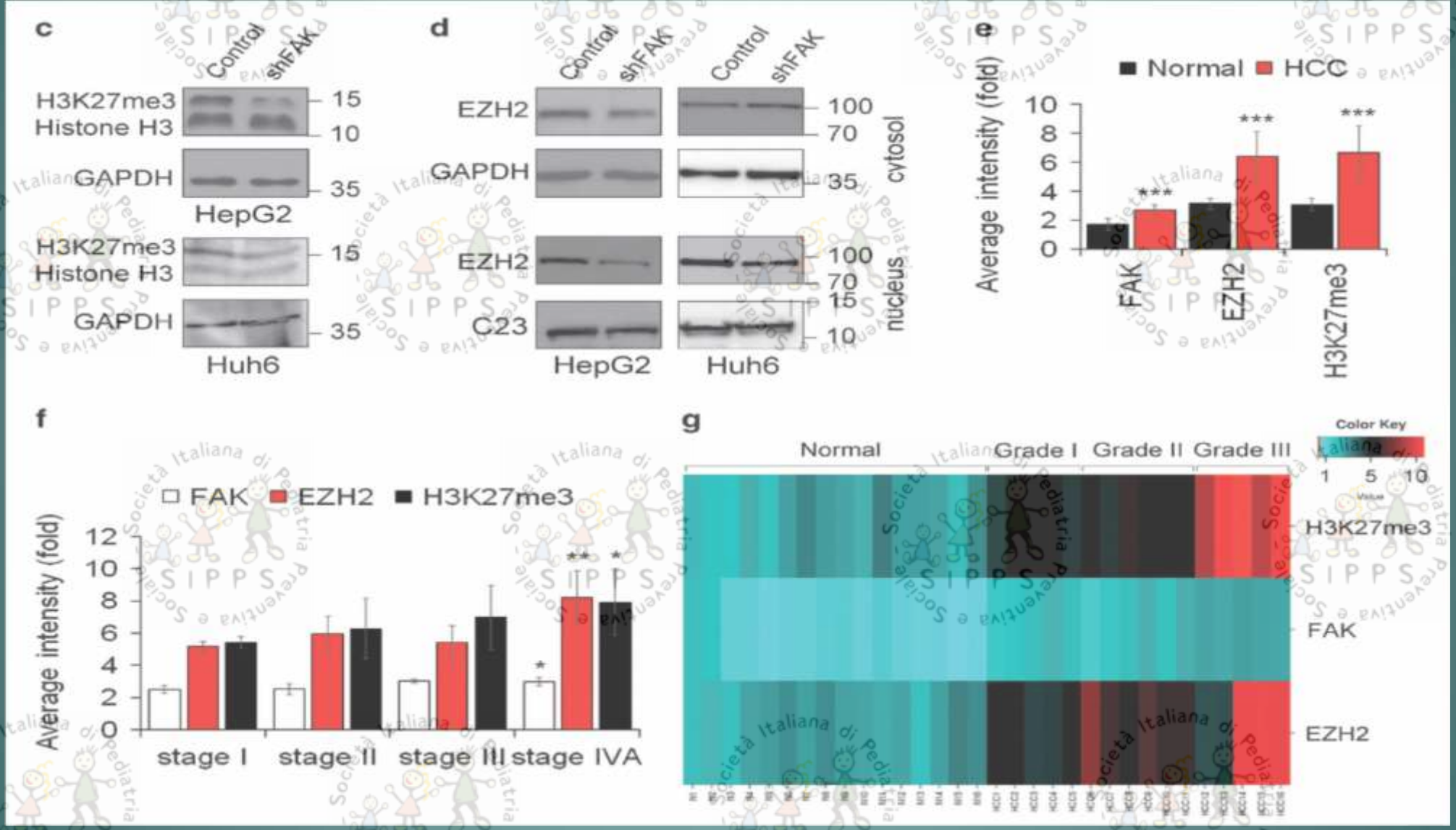
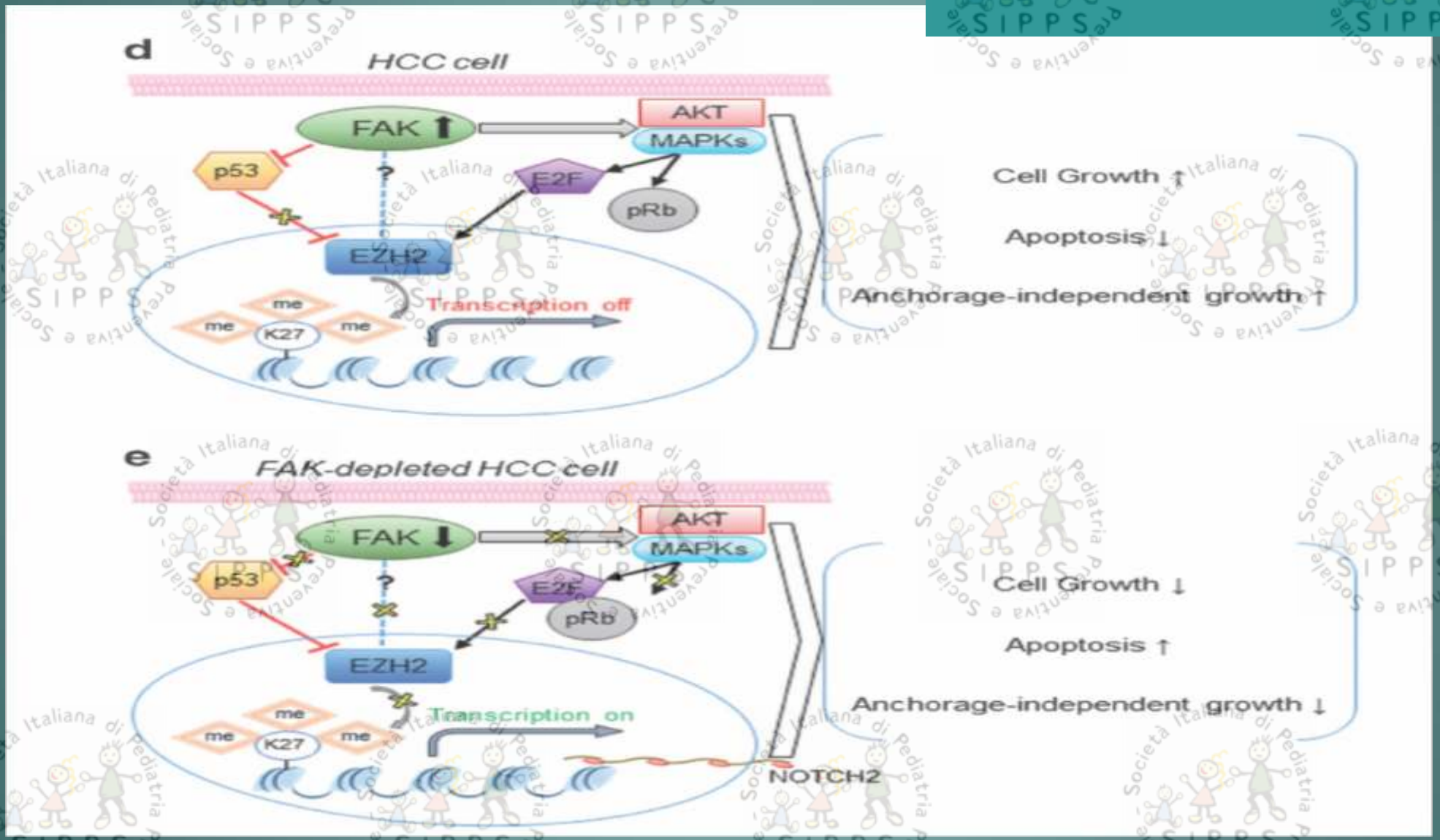
Max = 6.00e6


c
Control
shFAK
H3K27me3
Histone H3
15
10
GAPDH
35
HepG2
H3K27me3
Histone H3
15
GAPDH
35
Huh6
d
Control
shFAK
Control
shFAK
EZH2
100
70
GAPDH
35
cytosol
EZH2
100
70
C23
15
10
nucleus
HepG2
Huh6
e
Normal
HCC
Average intensity (fold)
10
8
6
4
2
0
***
***
***
FAK
EZH2
H3K27me3
f
FAK
EZH2
H3K27me3
Average intensity (fold)
12
10
8
6
4
2
0
**
*
*
stage I
stage II
stage III
stage IVA
g
Normal
Grade I
Grade II
Grade III
Color Key
1
5
10
Value
H3K27me3
FAK
EZH2


d
HCC cell
FAK
AKT
MAPKs
p53
?
E2F
pRb
EZH2
me
me
K27
me
Transcription off
Cell Growth ↑
Apoptosis ↓
Anchorage-independent growth ↑
e
FAK-depleted HCC cell
FAK
AKT
MAPKs
p53
?
E2F
pRb
EZH2
me
me
K27
me
Transcription on
NOTCH2
Cell Growth ↓
Apoptosis ↑
Anchorage-independent growth ↓

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Fructose increases risk of NAFLD in children

Intrahepatic innate immunity in chronic HBV infection

A comprehensive view on NS5A resistance

Abnormal IFN signaling in cirrhosis PBMCs

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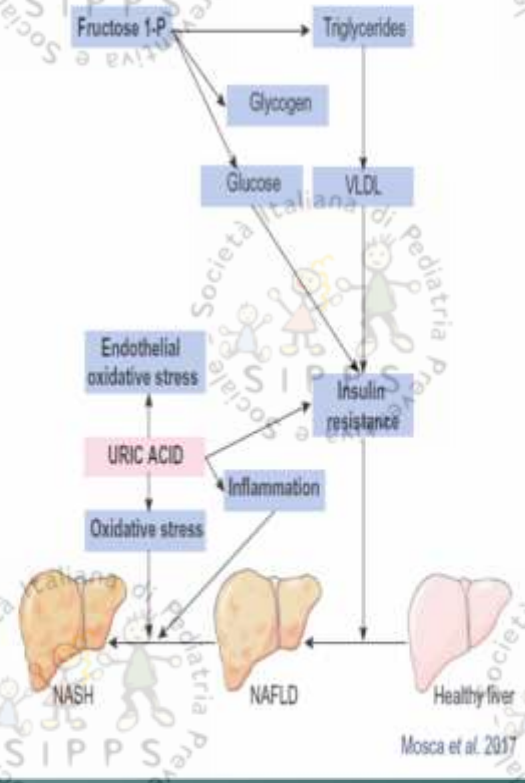
From the Editor's desk...

Richard Moreau*, Ramon Bataller, Thomas Berg, Jessica Zucmann-Rossi, Rajiv Jalan

SELECTION OF THE MONTH

Fructose consumption and hyperuricemia in young population

Excessive fructose intake is known to increase serum uric acid concentrations. In an important study by Mosca et al. a large cohort of children and adolescents with proven NAFLD (37% with NASH) were studied. Hyperuricemia was present in 47% of patients with NASH compared with 29% of non-NASH patients. Importantly, both **uric acid concentration and fructose consumption were independently associated with NASH**, after adjustment for multiple confounders. **Fructose consumption was the only factor independently associated with serum uric acid concentration.** This study suggests that excessive fructose consumption could lead to hyperuricemia and contribute to NASH development in young populations. Public health policies aimed at preventing excessive fructose-containing beverages among children are warranted.




Basta
bruciare
i grassi...
bruciamo i magri!
Destefano