Paventia z.s.

Český Krumlov

1st. INTERNATIONAL MEDICAL CONFERENCE ČESKÝ KRUMLOV

portal hypertension – Meso-Rex Bypass

in cooperation with

ISMETT Istituto di Ricovero e Cura a Carattere Scientifico

ČESKÝ KRUMLOV
April 05 - 07, 2018
Hotel BELLEVUE Český Krumlov

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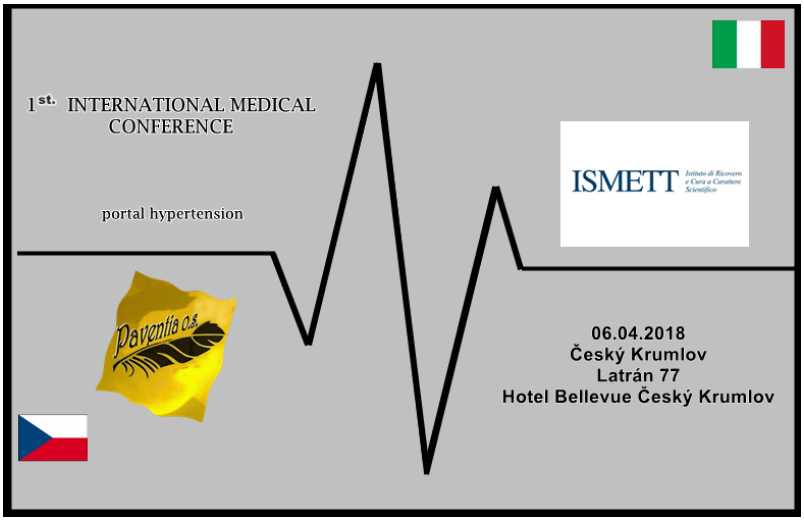
Prehepatic portal hypertension – Meso-Rex Bypass



April 05 - 07, 2018

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Český Krumlov
Therapeutic - educational center of the Paventia z.s.

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General information

Venue:

Hotel BELLEVUE Český Krumlov, Latrán 77, Czech Republic

Conference office:

the conference office will be set up for information and registration at the Paventia's training center a one day before the conference. On the day of the conference, the office will be at the front of the conference.

Thursady 05.04.2018 from 08:00 – to 15:00 o'clock in Paventia, Latrán 55, Český Krumlov
Friday 06.04.2018 from 09:00 in Hotel Bellevue Český Krumlov, Latrán 77, Český Krumlov

Official language:

Czech, English, Italian (simultaneous translation will be ensured)

Presentation time:

Lecture 15-20 min
Discussion 5-10 min

accommodation od participants:

Hotel Latrán Český Krumlov, Latrán 74,
Krumlovský mlýn, Široká 80 (sponzorováno - Marek Šimon)

WELCOME

Dear friends and colleagues,

I would like to welcome you to the first International Medical Conference in Český Krumlov dealing with portal hypertension and Meso-Rex-Bypass. We are proud to welcome participants from not only the Czech Republic but also from Italy - Sicily. We believe that this conference can start a long-term cooperation between medical practitioners in the Czech Republic and Italy - Sicily, especially with regards to the exchange of new information about various medical disciplines. This meeting will help us compare the achieved results, present our work and learn from each other.

Dear participants, take this event as an opportunity to get to know news in other medical disciplines that may be of a value to you and your work. Mutual overlapping of certain disciplines could result in significant benefits for all of us.

One important reason for organizing of this meeting is the potential and possibility for exchanging direct personal contacts. Nowadays, overloading information is a reality. You can easily find all the news, latest publications and scientific results on many websites. However, personal contacts open up space for useful discussions.

I thank all of you who have accepted the invitation to this conference because learning, scientific news and, above all, meeting with colleagues and friends will help to further our mission - to help people and especially children. If future mutual co-operation will have emerged from this meeting, this conference will have fulfilled its purpose.

I wish you a very fruitful scientific meeting and pleasant time in our beautiful city!

Mgr. Bc. Oldřich Hluško, Mgr. Marie Hlušková
PAVENTIA z.s., Český Krumlov
Therapeutic - educational center

PROGRAMME OVERVIEW

Friday – April 06, 2018

09:00 – 09:15	Meeting, registration	Discussion room
09:15 – 09:30	opening of the konference	Main lecture hall
09:30 – 10:10	Presentations n. 1	Main lecture hall
10:10 – 10:50	Presentations n. 2	Main lecture hall
10:50 – 11:05	Coffee Break	Discussion room
11:05 – 11:45	Presentations n. 3	Main lecture hall
12:00 – 12:45	Lunch	Golden hall
12:50 – 13:30	Presentations n. 4	Main lecture hall
13:30 – 14:10	Prezentations n. 5	Main lecture hall
14:10 – 14:25	Coffee Break	Discussion room
14:25 – 15:05	Presentations n. 6	Main lecture hall
15:05 – 15:40	Presentations n. 7	Main lecture hall
15:40 – 16:45	Discussion, Closing Ceremony	Main lecture hall

SCIENTIFIC PROGRAMME

Friday – April 06, 2018

09:30	**Štěpán Šembera M.D.**, Portal hypertension in the eyes of a gastroenterologist II. internal gastroenterology clinic of the Faculty Hospital Hradec Králové
10:10	**Pavel Piler M.D.,** Combined Transplantations – One Centre Experience Centre of Cardiovascular Surgery and Transplantation Brno
11:05	**Jitka Lukáčová M.D.,** Portal hypertension in the practice of a district pediatrician. Pediatrician in Český Krumlov
12:50	**Jean de Ville de Goyet MD**, PhD, FRCS., Meso-Rex Bypass – A Procedure to Cure Pre-Hepatic Portal Hypertension. Department of Pediatrics, ISMETT – UPMC, Palermo
13:30	**Maurizio Di Fresco M.D.,** International Patient Services at ISMETT International Patient Services Coordinator, ISMETT Hospital
14:25	**Michaela Lavičková, Mgr. et. Mgr., Ph.D.,** Payment of the health care services provided abroad by public health insurance The General Health Insurance Company, Regional office Plzeň, office for South Bohemian, Carlsbad and Pilsen district, director of law department.
15:05	**Mgr. Marie Hlušková,** Psychosocial effects of cavernom V. Portae chairman of Paventia z.s. Český Krumlov

Portal hypertension in the eyes of a gastroenterologist

Štěpán Šembera, M.D.
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Clinic of Internal Medicine and Gastroenterology II of the Teaching Hospital in Hradec Králové
Team of authors: T. Fejfar, V. Jirkovský, V. Chovanec, A. Krajina, M. Lojík, J. Raupach, O. Renc, P. Hůlek

Introduction:
Portal hypertension is overpressure in the blood vessels belonging to the portal vein (v.portae and its branches, v.lienalis, v. mesenterika superior and v.mesenterika inferior) and is defined by the porto-systemic gradient exceeding 5 mmHg. Clinically significant portal hypertension starts at 10 mmHg. Porto-systemic gradient cannot be measured otherwise than by catheterization of the jugular vein and follow-up wedging of the catheter or balloon in the jugular vein. Portal hypertension is classified based on the place where its cause is located into prehepatic, intrahepatic and post-hepatic.

Most frequently, portal hypertension results from hepatic cirrhosis where fibrotic changes in hepatic parenchyma affect the flow of blood through the liver – in this case intrahepatic portal hypertension (PH) is concerned. Other causes can be liver infiltration by tumour cells, sarcoidosis, congenital hepatic fibrosis, etc. Post-hepatic PH is mostly caused by Budd-Chiari syndrome, i.e. thrombotization of hepatic veins, or the adjacent inferior vena cava respectively. Budd-Chiari syndrome develops as a result of a procoagulation condition (mostly of a myeloproliferative disorder – polycythaemia vera, essential thrombocytemia, and myelofibrosis). Among other causes of post-hepatic PH is cardiac congestion. Among prehepatic causes of PH is thrombosis in the blood vessels of the portal vein. This can be induced by the same causes as Budd-Chiari syndrome, in addition with inflammatory diseases in the abdominal cavity (particularly pancreatitis, conditions after operations in the abdominal cavity), infiltration or oppression of veins by a tumour, in the paediatric population one of the frequent causes is the infection of the umbilical vein with propagation into the portal vein blood vessel system and formation of a thrombus. There are also other, rarer causes of portal hypertension.

Processes accompanying PH can be best explained in hepatic cirrhosis. In the case of fibrotization of liver tissue, individual sinusoids are impaired, resulting in an increased resistance to the flow of blood through the liver and therefore blood accumulates before hepatic parenchyma and congests. On the other hand, the increased pressure helps perfuse a more resistant organ and the cycle leads to an increased PH. In the case of overpressure in the portal system, porto-systemic collaterals develop, i.e. vein collaterals by-passing the liver into the systemic blood vessel system which, in a majority of cases, go through the oesophageal venous plexus into the v. azygos. Other places where blood can by-pass the liver with a lower pressure gradient is the left renal vein supplied from the lineal vein. Dilatation of these veins results in varices. Opening of portosystemic collaterals results in an increased flow through the portal vein blood vessels. Nevertheless, liver perfusion must be maintained even at the expense of increased effort, which means an increased cardiac output per minute. Increased perfusion pressure in the liver causes aggravation of lymph absorption and drain in the liver, which results in ascites.

The presence of varices in the oesophagus and stomach is associated with the risk of bleeding with up to 20% of lethal consequences. Ascites impairs the quality of life, can be infected by intestinal bacteria, can persist regardless of treatment (refractory ascites) and can be associated with hepatorenal failure. Among other complications of PH is hepatic encephalopathy, which is defined as a dysfunction of the central nervous system caused by hepatic insufficiency and/or portosystemic shunts and manifested by a wide range

of neurological and psychiatric abnormalities starting with clinically insignificant changes up to coma. Hepatic encephalopathy develops as changes in behaviour such as apathy, short temper, loosening of inhibitions, insomnia and disorientation. Another symptom is spleen enlargement with hypersplenism (consumption of blood elements, typically thrombocytopenia).

Prehepatic portal hypertension often manifests itself without any signs of liver impairment. The most frequent symptoms are varices and hypersplenism, while ascites is rare. Patients are endangered predominantly by bleeding from varices. On the other hand, with post-hepatic PH the liver is affected by ischemization and ascites is a frequent sign; varices and bleeding from them is less frequent.

Budd-Chiari syndrome can have an acute up to a chronic form. With the acute form, there is a risk of hepatic failure due to hepatocyte ischemia. With the chronic form, hepatic cirrhosis develops due to congestion, which manifests itself as chronic liver failure. Thrombosis in the portal vein blood vessel system can also be acute, in which case it manifests itself by abdominal pain with a risk of infarction of the intestine with vascular ileus, or chronic, when a number of collaterals develop in the abdominal cavity that impose as a cavernoma. Such a case is called cavernous transformation of the portal vein. Patients are endangered by bleeding varices.

If possible, treatment should focus on the causes of portal hypertension (in the case of hepatic cirrhosis, patients should not drink alcohol; viral hepatitis should be treated, etc.). In the case of Budd-Chiari syndrome and acute forms of thrombosis in the portal vein anticoagulants should be administered. If larger varices are ascertained or in the case that the patient has experienced an episode of bleeding, a non-selective beta blocker (e.g. Carvedilol) should be administered and it is also important for the patient to undergo an endoscopic treatment of varices, usually by ligation. With ascites, diuretics are administered, in the case of secondary hyperaldosteronism, aldosterone antagonist – Spironolakton, is preferred, followed by Furosemide. If ascites fails to recede, regardless of a gradual increase in the dose of diuretics up to the maximum dose, paracentesis needs to be performed repeatedly. Another option is to implant a transjugular intrahepatic portosystemic shunt (TIPS), i.e. a stent between the portal vein and the hepatic vein. A TIPS is indicated in the case of bleeding that cannot be stopped endoscopically with ineffective prevention of bleeding (the patient has recurring episodes of bleeding regardless of ligation and treatment by beta blocker) and in the case of refractory ascites. A disadvantage of a TIPS is an increased risk of hepatic encephalopathy. A TIPS has nearly replaced surgically created portosystemic shunts. Nevertheless, in certain cases, in particular with prehepatic portal hypertension where TIPS is usually not regarded as a suitable treatment, a surgical solution is necessary. In addition to portosystemic shunts, splenectomy with azygo-portal disconnection (interruption of collaterals in the wall of the stomach and distal oesophagus) is available. In clinically serious cases that cannot be treated alternatively, transplantation of the liver should be considered. We submit experience from the Teaching Hospital in Hradec Králové (FNHK) with the treatment of PH.

Methodology:
The group of patients includes all patients identified at the Clinic of Internal Medicine and Gastroenterology II of the FNHK with acute or chronic thrombosis and obstruction in the portal vein blood vessel system that are directly caused by a malignant tumour or cirrhosis. The retrospective description of the group includes the following items: demographic parameters, the aetiology of the obstruction of the portal vein blood vessel system, classification into acute and chronic obstructions, manifestations, and therapeutic procedures.

The results:
The group includes 52 patients, of whom 27 are males and 25 are females, 44 patients with chronic cirrhosis, 6 patients with acute thrombosis and 2 patients with subacute thrombosis.

The aetiology of the portal vein blood vessel system obstruction was as follows: coagulopathy (Leiden mutation, deficit of the C and S proteins, deficiency of antithrombin III, antiphospholipid syndrome and others) was discovered in 13 patients, a myeloproliferative disorder was ascertained in 8 patients, and 6 patients had either acute or chronic pancreatitis. Within the framework of therapy, 12 patients underwent either acute or elective splenectomy, 7 patients were performed an azygo-portal disconnection, 6 patients were implanted with a splenorenal shunt and 10 patients received a TIPS. In the case of acute thromboses, local thrombolysis was performed in 6 cases. Portal biliopathy with portal cavernoma was observed in 3 cases.

Conclusion:
Acute and chronic thrombosis of the portal vein in patients without cancer or cirrhosis is a clinically serious and life-threatening condition requiring various endoscopic, radiological and surgical interventions. Only a part of these approaches supports the currently recommended procedure.

References:
HŮLEK, P.; URBÁNEK, P., Hepatologie 2018. Prague: Grada, 2018. In the process of publication. Chapters 5.3, 16

European Association for the Study of the Liver, EASL Clinical Practice Guidelines: Vascular Diseases of the Liver. Journal of Hepatology 64, no. 1 (January 2016): 179–202

Combined Transplantations – One Centre Experience

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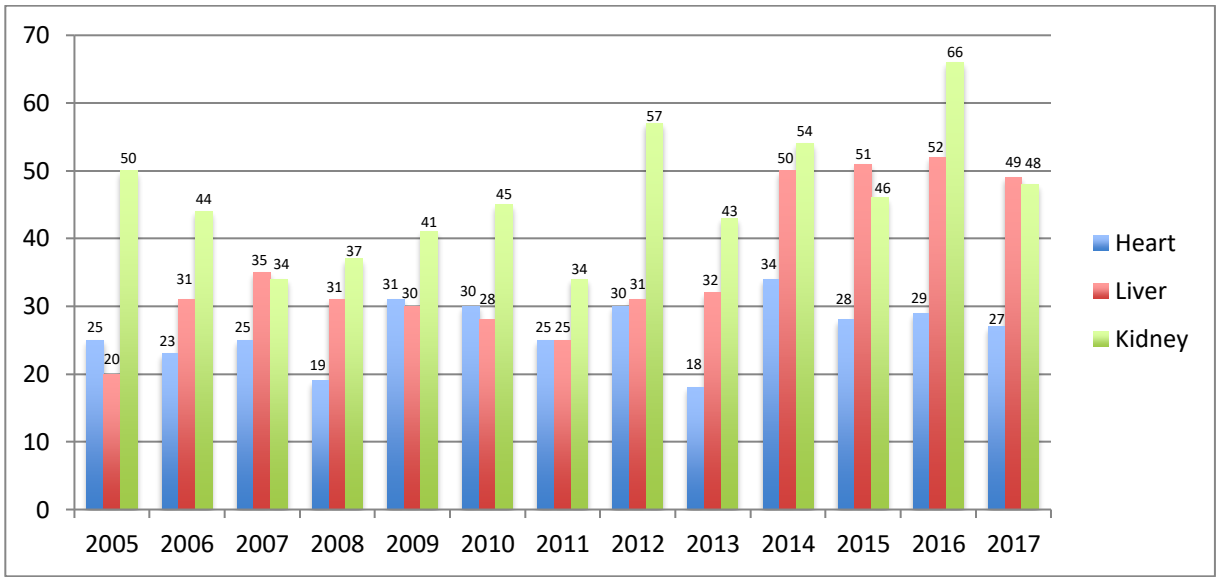
Centre of Cardiovascular Surgery and Transplantation Brno, Consultant for cardiac surgery and transplantation
Co-authors: Jiří Ničovský, M.D.

Introduction and Aims

This brief communication informs about experience with combined organ transplantation. In Centre of Cardiovascular Surgery and Transplantation (CKTCH) in Brno, a complete spectrum of cardiac surgery, heart transplantations, liver transplantations and kidney transplantations is performed. This fact creates the best base for combined organ transplantations. Numbers of performed organ transplantations are visible in Graph No.1.



Graph No. 1: Numbers of Transplantations 2005 – 2017

The aim of the Communication is to inform about experiences with combined transplantations performed in CKTCH Brno from 2005 to present. In detail, the report deals with concrete results of two parts, first part, where heart and kidney are transplanted at the same time simultaneously and the second part, where the kidney was transplanted after heart transplantation within a time delay.

Methods

In the first section authors analyse the case report regarding triple transplantation - simultaneous transplantation of heart, liver and kidney of one patient. Surgery, perioperative data and results are pointed.

In the next section authors describe results of combined transplantations in detail. Numbers of combined transplantations are in Table No. 1. Combined transplantation of liver and kidney is analyzed separately. The third section informs about combined transplantation of heart and kidney in two parts. First part, where heart and kidney are transplanted at the same time simultaneously and the second part, where the kidney was transplanted after heart transplantation consecutively, with delay. Reasons of heart and kidney failures are analyzed and there are also included demographical and clinical data, operational data and postoperative data. In the consecutively transplanted kidney cases there are also mentioned a time distances between heart transplantation and consecutive kidney transplantation.

	Simultaneously	**Consecutively**	**Total**
Heart + kidney	8	13	21
Liver + kidney	22	8	30
Heart + liver + kidney	1	0	1
Total	**31**	**21**	**52**

Table No. 1: Numbers of simultaneously and consecutively transplanted organs

Results

Authors informs about mortality of heart, liver and kidney transplantations, of combined liver and kidney transplantations and of heart and kidney transplantations in two parts – simultaneous and consecutive transplantations.

Simultaneous transplantations – heart + kidney

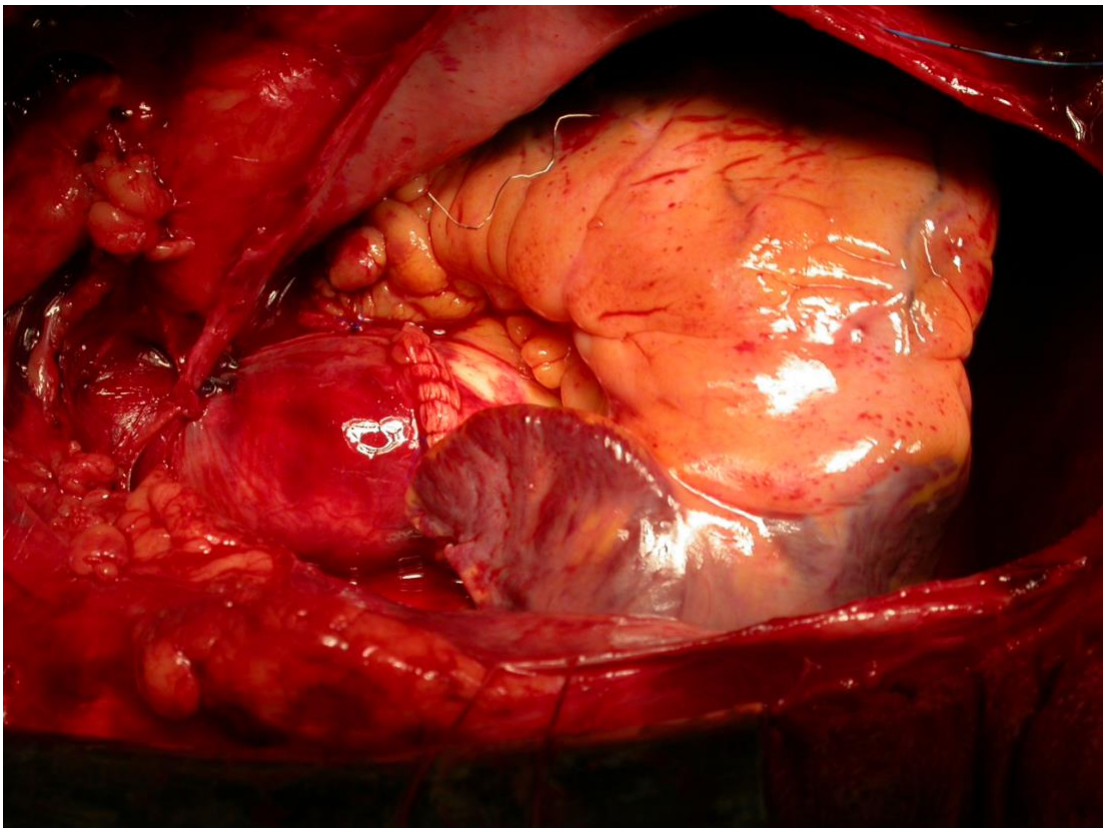
- 6 alive
- Death 15 years post OP (heart failure)
- Death 2,5 years post OP (brain bleeding)

Consecutive transplantations – heart + kidney

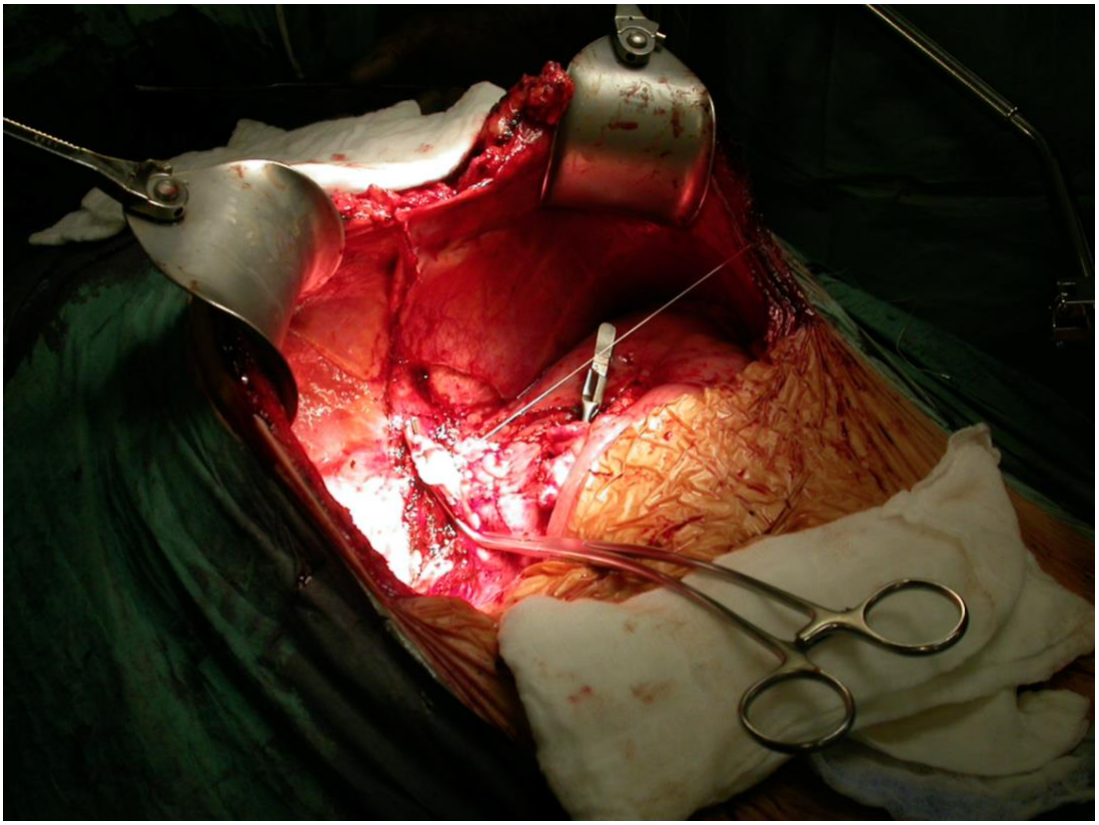
- 10 alive
- Death 3,5 years post OP (MOF)
- Death 3,5 years post OP (GIT bleeding)
- Death 20,5 years post OP (craniotrauma)

After triple transplantation was patient alive 2,5 years and died because of intracranial bleeding.

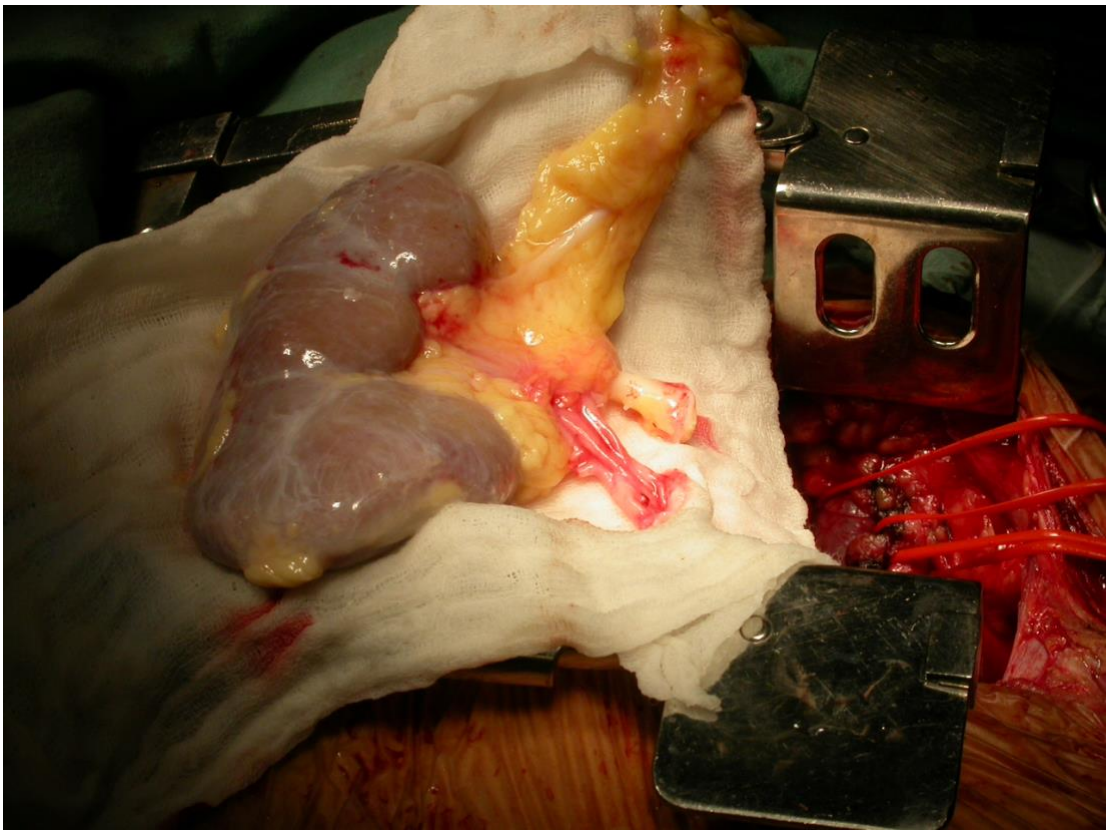
At pictures it is possible to see results of triple transplantation – transplantation of heart (Picture No. 1), transplantation of liver (Picture No. 2) and transplantation of kidney (Picture No. 3). Picture No. 4 covers all three surgery areas during triple transplantation.



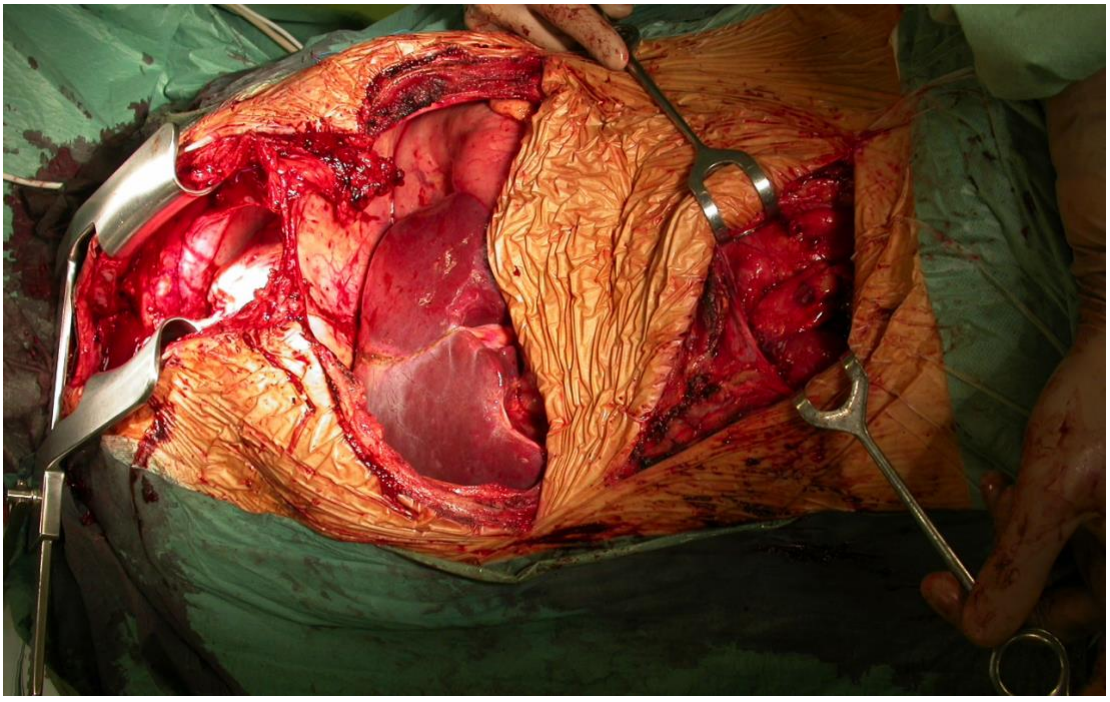
Picture No. 1: Heart transplantation



Picture No. 2: Liver transplantation



Picture No. 3: Kidney transplantation



Picture No. 4: Triple transplantation

Conclusions and Summary

Based on presented data it is possible to declare that combined transplantations are an appropriate and safe method for patients with multiple organ failure which require transplantation of several organs. Mortality of these patients is not significantly different from mortality of single organ transplantations. But the number of combined transplantations is very low, combined transplantations are rare.

These procedures can be performed only at a centre where is extensive experience with transplantations of organs and where the frequency of these transplantations is high and results are appropriate.

Essential and basic requirements are adequate personal staffing, diagnostic capabilities, perfect equipment for procedures and perioperative monitoring including modern applications (ECC, ECMO, CVVHD, CT, MR, ...). Necessary element is patient monitoring and post-operative complications care.

Absolutely essential for this process is team-cooperation and perfect collaboration between surgeons, anesthesiologists and intensivists, cardiologists, hepatologists and nephrologists, and perfect complement base.

Portal hypertension in the medical practice of a paediatrician (2 case studies)

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Pediatrician in Český Krumlov

PORTAL VEIN OBSTRUCTION

Portal vein obstructions at an early age are rare. They are diagnosed usually due to coincidentally discovered thrombocytopenia, splenomegaly or bleeding from oesophageal varices. This is given by permanently increased blood pressure in the blood vessels belonging to the portal vein. Causes: Portal hypertension can have many causes that are divided into three basic groups depending on localization.

Prehepatic causes (congenital anomaly of the portal vein and its blood vessel system, infectious disease in the abdominal cavity – for example peritonitis, rarely appendicitis, cholecystitis, cancer in the abdominal cavity, and thrombosis of the portal vein), hepatic causes (hepatic cirrhosis, hepatic fibrosis, blood diseases – leukaemia, lymphoma, toxic liver failure, and cancers) and post-hepatic causes such as a tumour growing through the portal vein, thrombosis (formation of a blood clot) and right-sided heart failure.

Symptoms and signs of portal hypertension: oesophageal varices, splenomegaly, or possibly hepatomegaly, ascites, fatigue, loss of appetite, constipation, flatulence, low sexual desire, pressure and pain in the hypochondrium and abdominal cavity, loss of body weight. Further: spider naevi, excessive bleeding, impairment of the cerebral functions (encephalopathy).

Determination of the diagnosis of portal hypertension: ultrasonic examination of the abdominal cavity, CT, MRI angiography, pressure measurement in the portal vein: catheterization of hepatic veins. Blood count (thrombocytopenia), clinical blood chemistry (content of proteins, ions, blood coagulability).
Treatment of portal hypertension

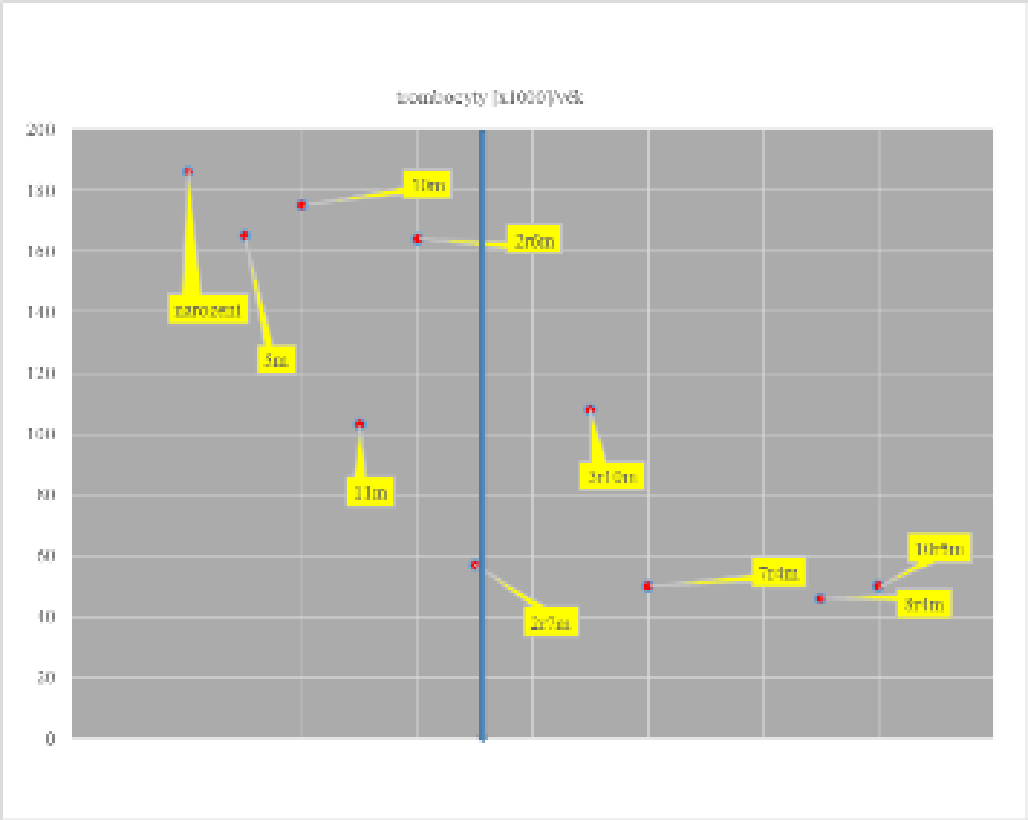
Treatment of oesophageal varices and bleeding from them (most frequently by endoscopic ligation or endoscopic sclerotherapy.
Reduction of pressure in blood vessels belonging to the portal vein using vascular stimulants. For example, vassopressin is administered.
In the case of massive bleeding, urgent surgery on the oesophagus can be performed if the previous procedures failed.

A transjugular intrahepatic portosystemic shunt (TIPS) can be implanted where the intervention radiologist uses a stent to connect the portal vein and the hepatic vein through the liver.

CASE STUDIES

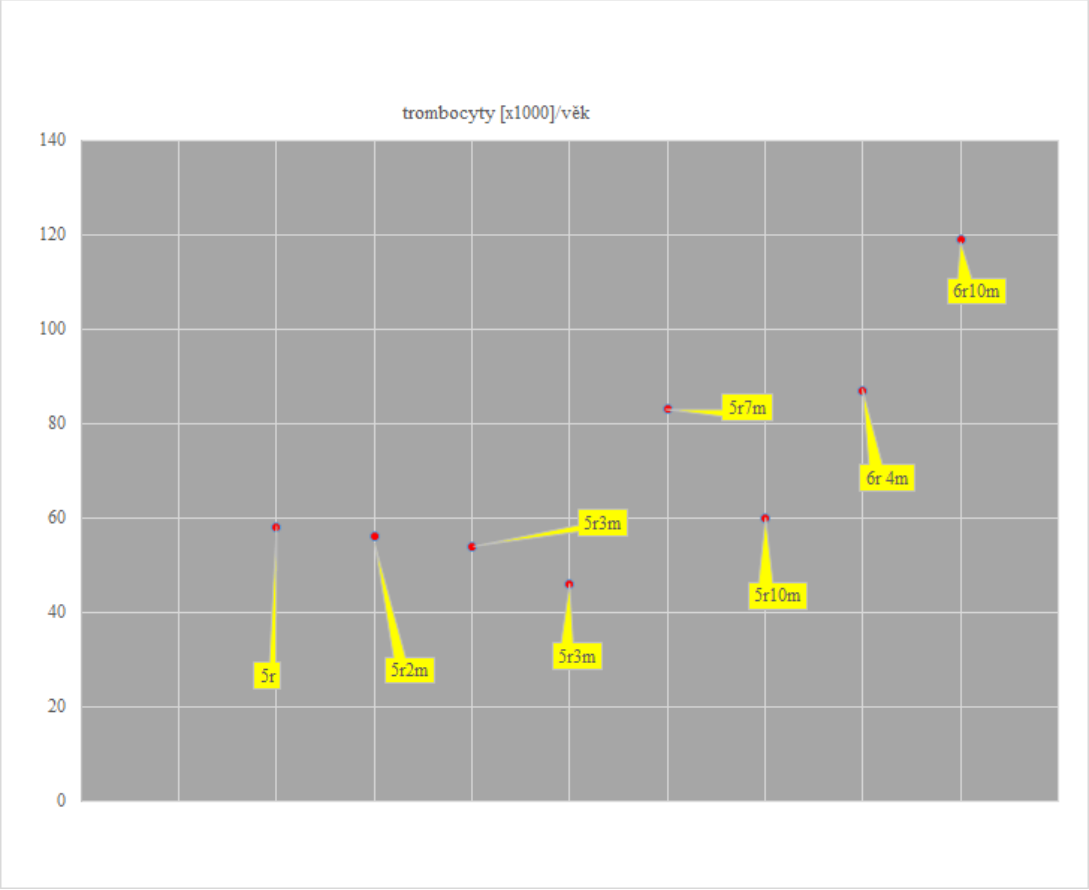
Case No. 1:

A ten-year-old boy born as a second child. Delivery by Caesarean section due to imminent hypoxia. Birth weight/birth length 3,700 g/51 cm, due to a postpartum breathing disorder with turbid amniotic fluid, he was transported to the Neonatology Department in the hospital in České Budějovice where the cannulation of the umbilical vein was performed because of incipient sepsis with antibiotics being administered. He was hospitalized for 10 days and released in good condition. Up to May 2010, his blood count was tested a total of four times for various reasons, including thrombocytes, always with normal findings. In April 2007 he went through varicella after which he was tired and therefore his blood count was examined with discovered anaemia and thrombocytopenia (HB 107 g/l, Ht 32.3%, thrombo 57 thous.). As splenomegaly was discovered, he was sent to the outpatient department of the paediatric clinic in České Budějovice where medium thrombocytopenia was ascertained with 89 thous./l. However, after a week the thrombocytopenia was reduced to 158 G/l. An ultrasonic examination discovered splenomegaly and portal hypertension. Portal hypertension after an episode of perinatal infection and catheterization of the naval was the conclusion. The child was handed over for observation by a paediatrician – gastroenterologist. In April 2012 endoscopic oesophagogastroduodenoscopy was performed which ascertained oesophageal varices dilated up to 5 to 7 mm. Another follow-up EGD examination was performed two years later, the last time in October 2016 and another one is scheduled in autumn this year. The boy is monitored by the gastroenterologist and haematologist. He is rarely ill, without subjective difficulties and the only objective finding is splenomegaly ascertained by ultrasonic examination 216×84×142 mm, the count of thrombocytes in March 2018 was 50 thousand per cm^3.



Case No. 2:

A seven-year-old girl born as a fifth child, high-risk pregnancy due to IVF and previous abortions. Delivery by Caesarean section due to the foetal breech position in the 39th week of pregnancy. Birth weight 2,900 g/47 cm. Postpartum adaptation good. In March 2013, she was operated on for a left-sided inguinal hernia. Otherwise she was not ill. When she was five, it was discovered that her spleen was enlarged. Due to thrombocytopenia 58 thousand and splenomegaly in July 2016, sternal tapping gradually excluded a haematological disease as well as storage metabolic disorders at the Clinic of Paediatrics in Motol University Hospital and in Karlov. In September 2016, MRI and MRA of the liver and abdominal cavity were performed with cavernous transformation of the vena portae ascertained in the porta hepatis region, wider in mes. sup. and vena lienalis, with retrogastritic, perisplenic and splenorenal portosystemic collaterals as a symptom of portal hypertension and a finding of splenomegaly. In October 2016, an EGD examination was performed with one ascertained oesophageal varix in the length of 4 cm above the cardia. The girl suffered from significant splenomegaly (+13 cm) and thrombocytopenia (40 to 50 thousand). At that time, her parents contacted a clinic in Palermo and decided on Meso-Rex bypass. In October 2017 the girl underwent the operation and recovered without any major complications. One month after the operation she had 87 thousand thrombocytes and in January 2018 her thrombocyte count reached 119 thousand. She is in good condition and experienced no serious illness.



Meso-Rex Bypass – A Procedure to Cure Pre-Hepatic Portal Hypertension.

Jean de Ville de Goyet MD, PhD, FRCS.

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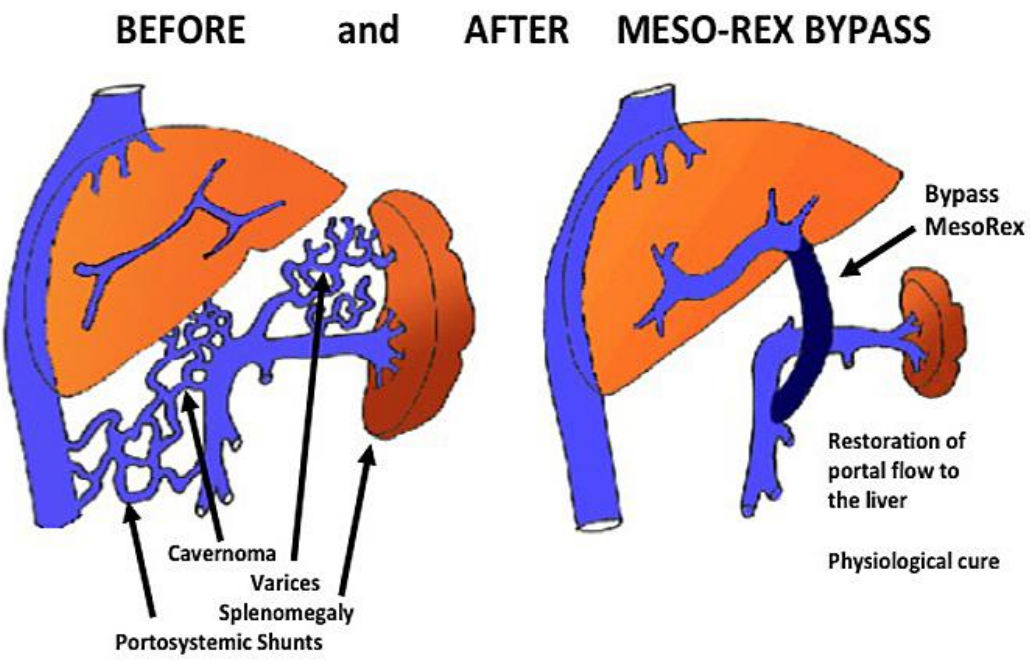
ISMETT – UPMC
Via Ernesto Tricomi 5, 90127 Palermo, Italy

Pre-hepatic portal hypertension is a condition typically characterized by the thrombosis of the portal vein in presence of a healthy liver. This condition also evolves typically with the development of a "cavernoma" in the place of the portal vein (numerous venous collaterals developing in an anarchic manner). Pre-hepatic portal hypertension with cavernomatous transformation of the porta hepatis is the single most common cause of portal hypertension in children. It can be secondary to congenital thrombophilia or a neonatal catheterization of the umbilical vein (25% of cases), or secondary to trauma, infection, inflammation or regional surgery. However, in most cases, no etiological cause is found ("idiopathic" cavernoma).

The Meso-Rex Bypass was first described in1992. In a few words, this procedure consists of bypassing the thrombosed portal trunk (and the cavernoma), by interposing a graft between the superior mesenteric vein and the recessus of Rex (a portion of the left portal venous system). With time, the technique has gained interest, and many teams around the world started implementing this original approach into their clinical practice; good outcomes have been reported repetitively, the major advantage of the procedure being a natural and physiological cure (by restoring the hepatopetal portal flow) of portal hypertension, to the contrary of conventional porto-systemic shunts that are palliative procedures, as they treat portal hypertension by diverting the splanchnic blood flow away from the hepatic route.

The Meso-Rex Bypass reduces portal pressure by reopening an access to the low-resistance hepatic parenchyma. In turn, the Meso-Rex Bypass allows the regression of natural porto-systemic connections, and reverses, if any, the side-effects related to either portal hypertension and/or porto-systemic connections. It has been shown that it reversed hepatopulmnary syndrom, and that it improves neuro-cognitive ability, helps normalizing hyperammonemia, or reverses encephalopathy caused by porto-systemic connections. MRB may prevent the formation of liver nodules and adenoma in children, with natural or surgical porto-systemic shunts, and reverses the commonly observed coagulopathy in patients with cavernoma (elevated prothrombin time and INR, with decreased levels of factors V, VII, protein C and protein S. Lastly, it improves somatic growth in those patients who have displayed growth retardation.

The Meso-Rex Bypass was recently recommended as being the first choice of surgery for children with extrahepatic portal hypertension, a healthy liver and a patent Rex recessus. Some recommend to proposed the procedure rather early in life rather than postpone it to adult age - as often done for conventional portosystemic shunts- and proceed as a pre-emptive management; this strategy helps preventing many of the portal hypertension co-morbidities or complications that develop with time and the growing of age.



In conclusion, the Meso-Rex Bypass is a real « cure » of extra-hepatic portal hypertension, because of its ability to reestablish physiological hepatic portal venous blood flow into and through the liver. This is clearly advantageous over traditional palliative methods and portosystemic shunting. Because it is a physiological cure, it has evolved to become nowadays the gold standard in the management of extra-hepatic portal hypertension in children with a healthy liver and a patent Rex recessus.

Beneficial Effects of the cure of portal cavernoma by a Meso-Rex Bypass

Year – Author (reference)	**Beneficial Effect Type**	**Patient Number**
1992, de Ville de Goyet (22)	Index case: Cure of portal hypertension after liver transplantation	1
1998, de Ville de Goyet (26) 2006, Superina (35) 2010, Sharif (34) 2013, Lautz (33)	Physiological Cure of Portal Hypertension in children with Idiopathic Pre-Hepatic Portal Hypertension (larger series)	7 31 30 65
2003, Gherke (29)	Restores the Hepatopetal Portal Flow and Intra-Hepatic Portal System Flows	13
2003, Mack (13)	Reverses the Commonly Observed Coagulopathy	14
2013, Lautz (33)	Normalizes Hyperammonemia	65
2006, Mack (37)	Improves Neuro-Cognitive Ability	12
2006, Chui (38)	Reverses Encephalopathy caused by Porto-Systemic Connection	1
2007, Stringer (39) 2009, Lautz (40)	Improves Somatic Growth in those Patients who had Growth Retardation	11 45
2003, Fuchs (30)	Reversed Hepatopulmnary Syndrome	1
2009, Guerin (20)	Prevents Formation of Liver Nodules and Adenoma related to Porto-Systemic Shunting	1 (personal communication)

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International Patient Services at ISMETT

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ISMETT is a scientific-based care and research hospital, located in Palermo, Italy, active in the field of end-stage organ failure. A state-of-the-art transplant center and a reference hospital for the entire Mediterranean area, ISMETT is involved in important research projects designed to provide patients with the most advanced therapies for end-stage vital organ failure.

An example of innovative and efficient clinical management, ISMETT was created through an international partnership between the Region of Sicily and UPMC (University of Pittsburgh Medical Center).

The collaboration with UPMC, a leading institution in the field of organ transplantation, brought to Palermo the expertise and professionalism of the American worldwide-renowned hospitals, research centers, and universities.

ISMETT is the first hospital in southern Italy to have received the Joint Commission International (JCI) accreditation, which is considered one of the most advanced accreditation systems for assessing the quality and safety of hospital facilities. JCI accreditation confirms ISMETT's top standards of patient care and patient safety, and its commitment to ongoing improvement of clinical outcomes, management, reception, and training programs that involve many hundreds of nurses, physicians, technicians, and administrative employees every year.

ISMETT's activity focuses on the treatment of patients with complex diseases, including but not limited to transplant programs, which are often the most appropriate therapeutic option or irreversible cases. Currently, ISMETT offers all solid organ transplant programs: liver, kidney, heart, lung, and pancreas for both adult and pediatric patients.

Transplantation is not the only answer to end-stage organ failure. A certain number of these cases can be successfully treated with medical therapies, surgeries, and interventional radiology, endoscopy and cardiology procedures. ISMETT's competences are available also for patients who do not need transplantation, but suffer from complex diseases.
Whenever possible, minimally invasive approaches are chosen, to the benefit of the patients.

The tables below show:

Table 1: trend of foreign patients hospitalizations at ISMETT from 2005 to 2017

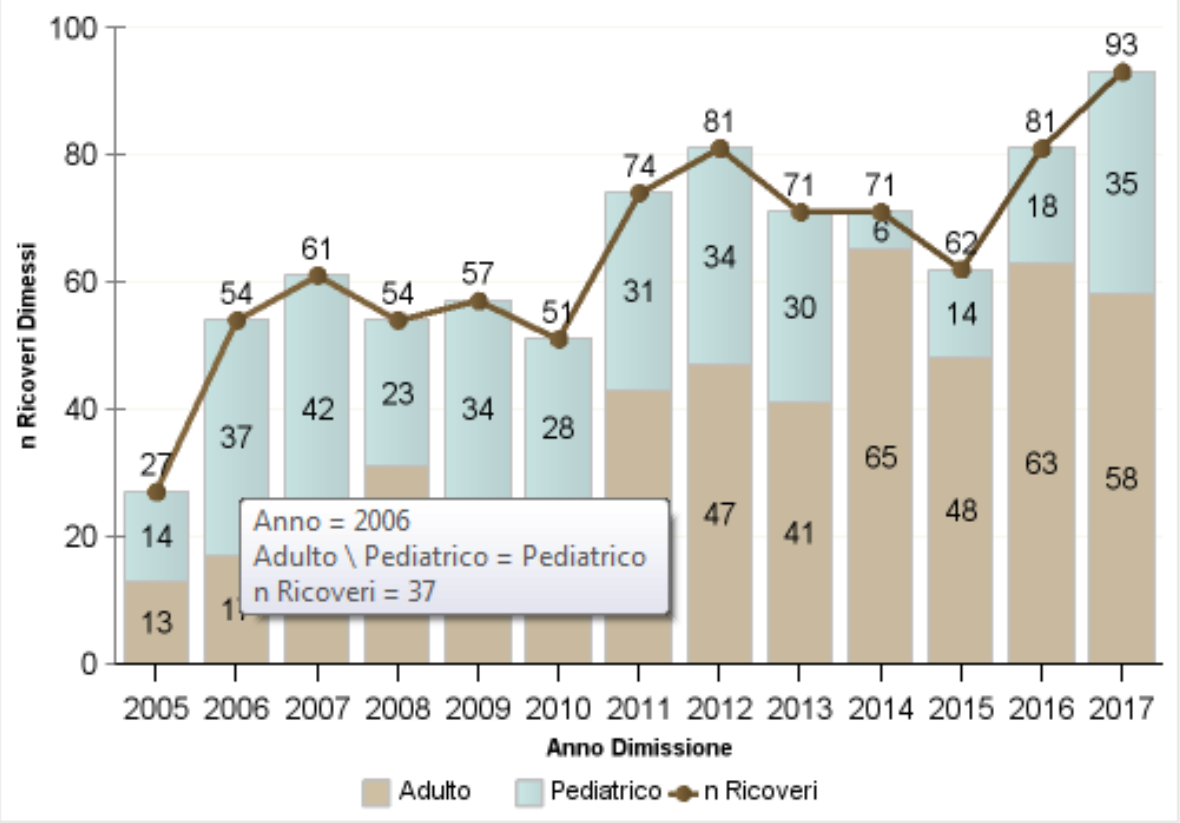


Table 2: categories of foreign patients at ISMETT from 2004 to 2016

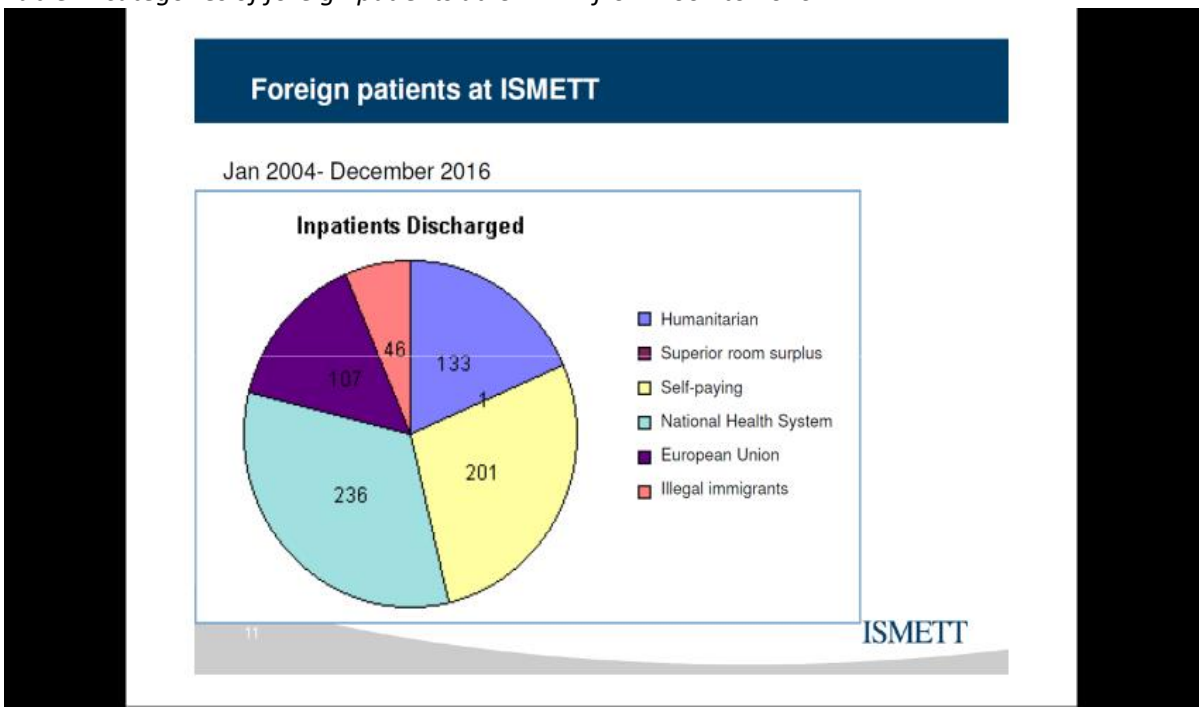
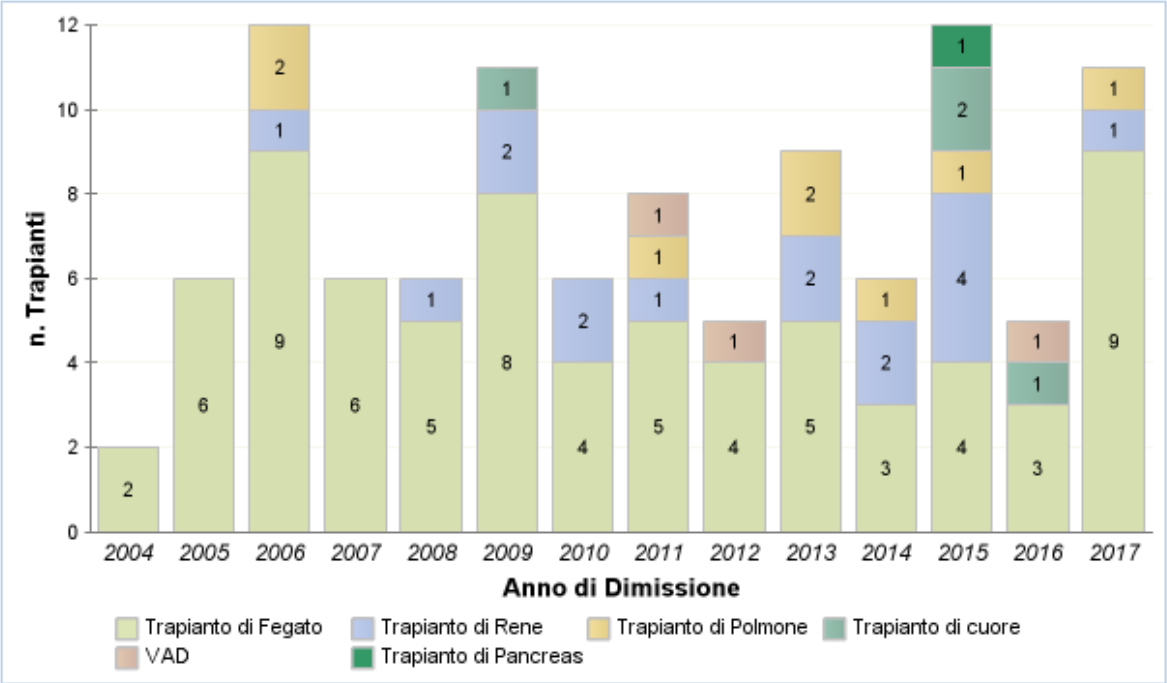


Table 2: organ transplants carried out on foreign patients



The international Patient Services (IPS) Department of ISMETT provides the highest level of personalized and culturally sensitive services to patients seeking outstanding medical care and warm hospitality. Whether seeking an initial diagnosis, a treatment for a diagnosed disease, or a periodic check-up, patients benefit of a comprehensive approach from a professional team, and each patient receives the highest level of care and comfort.

The IPS coordinator's support will continue also during the follow-up after the treatment is complete and the patient has returned home. By performing a variety of services, our team strives to handle all logistics, so patients are free to focus on their treatment and healing.

In the adult department, dedicated suites are available that offer attentive, quality care in a private and comfortable atmosphere. Each one of such dedicated rooms boosts a foyer equipped with a microwave, a refrigerator and other kitchen supplies, and a main area with a foldout couch for one guest, a bathroom, a strong box and complete in-room entertainment including satellite TV (allowing for a wide range of foreign channels), computer, Internet access, telephone, fax, printer, play station, DVD player.

One family member is always permanently allowed inside our International Patient Suites, with meals included also for the accompanying person.

The International Patient Services Department guarantees a dedicated attention to the peculiar needs of people coming from other Countries.

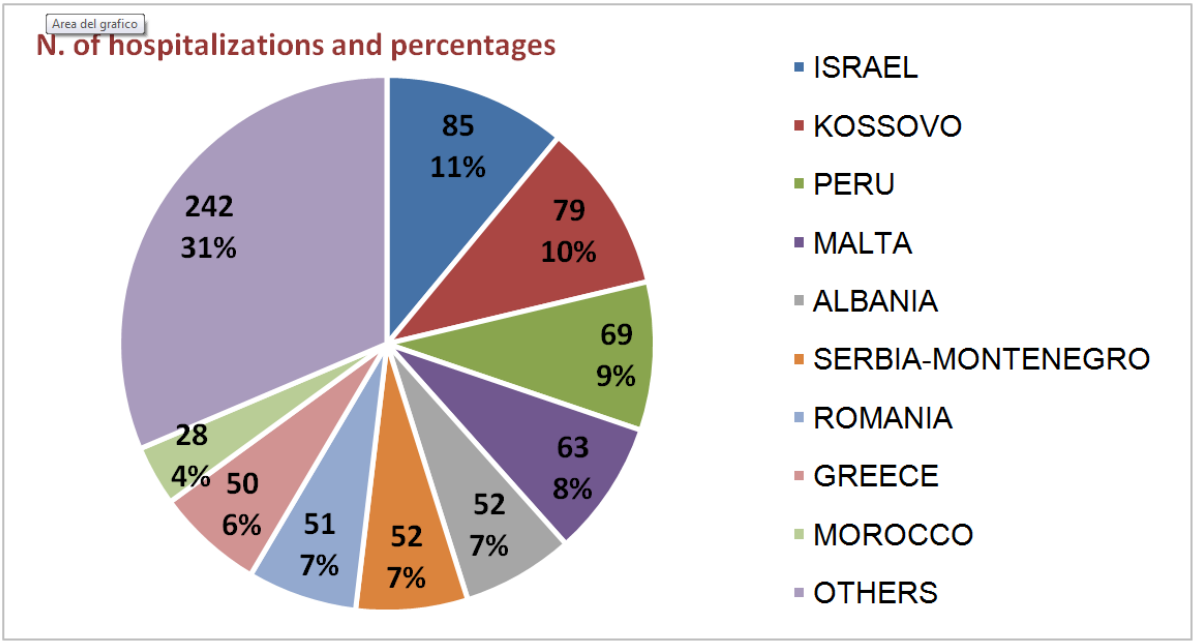
From the first inquiry until the return home, each patient is followed through all stages of care by a specialized team, and is offered personalized services. The most scrupulous attention is paid to the quality of medical services and to the needs of patients and their families.

The International Patient Services Department offers assistance to international patients before, during and after their stay in Palermo, with services that include:

- Administrative and financial services (assistance with hospital admission procedures, doctors' appointments, financial transactions, cost estimates and billing, contact with embassies and consular offices, visa requests/extensions)

- language services (translation of medical documents into English for use upon return home, native language interpreter's presence during hospital stay)
- Accessory services (help in finding accommodations, transportation assistance, special foods and nutritional needs, airport pick-up service, air ambulance, rehabilitation, pharmaceuticals, medical devices, cultural and religious dietary needs, assistance with religious needs).

Divided by Countries of origins, the figures hereafter summarize the main places where foreign patients have come from during the period 2005-2017:



Thanks to the inauguration of a new Department of Pediatrics in October 2017, which has expanded the number of beds, and with several development projects to be implemented in the next few months, ISMETT's foreign patient population is growing steadily, with particular regard to patients coming from all over Europe, as new therapeutic and logistic options are being and will be added to the International Patient Services, always valuing each and every need of all patients.

Payment of the health care services provided abroad by public health insurance

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Introduction
The payment of the health care services provided abroad by public health insurance differs according to the form of care provided. So if the care is ambulant or hospital, planned or accidental, in some case urgent or pressing. Significant is also, in which country outdoors, the care is provided.

Discussion
According to the Czech law 48/1997, about public health insurance, are from the public health care system covered the services provided the insured person which lead to the improvement or maintenance of his/her state of health or reduce his/her suffering. Further, these services have to come up with the health and purpose needed and are appropriately safe, consistent with the recent available finding of the medical science and there is evidence of its efficiencies. These are mandatory cumulative conditions.

First of all these covered health services are to be provided in Czech Republic. The law do not exclude their providing for the people participating in the Czech public health insurance program also abroad. Primarily the situations when the need of providing this care during the staying abroad is thought.
Extension of the covered health care according to the EU law:
1) medical necessary health care services
- the insured person, staying abroad but in EU temporarily must get the health care services in the extension (according to the intended length of stay), the patient must not travel to the insurance country before he/she intended.
2) health care services are provided in the full extend according to the particular state law
- the insured person, resident and registered in a state different from the insurance state. This person has the claim on the same care, on which has the claim of the local insured person.
3) requested planned health care services
- specific care, the patient travel abroad to target, due to already existing disease or health problem, health care. The claims on this care have the people who obtained the acceptance of the insurance company with the payment of planned and provided health care services abroad. In case of ambulant care the previous acceptance of the provided care from insurance company is not being asked and the person get, after submission all the documents needed (usually confirmation of provided services and the payment) refund of expend according to the Czech law. But in the event of planned hospital care, the previous acceptance of insurance company is always demanded.

The basic criteria for approving covering of the planned hospital care in Czech system insured person is especially unavailability of this care in Czech Republic. This care also must be paid by Czech public health insurance system. The request should not be also denied if the care is provided in Czech Republic but the health care services cannot be provide in “medically justificative period”. It means that the view point of accessible care is not only the fact the care is in the Czech Republic provided but also if the care the patient can get within certain time which in some cases can be the most crucial aspect. Under consideration is thus the access, seasonableness and actual state of health and expected development of the disease. So the very individual consideration is always needed. The application for this care should be handle within 30 days, according to the Czech law.

There are some exceptions or specificity from that mentioned above. The Czech public health care system is relatively wide and the patients are asked to make a co-payment towards the cost very seldom, usually only towards the cost of certain drugs. During hospitalization, an insured person does not pay any cost. In other state, also within EU, is not rare, that the specific health care in Czech Republic covered, in different public health system are uncovered or co-paid. The acceptance of the insurance company with providing health care abroad is also valid only for the facilities involved in public health care system in concrete state. Without the document of acceptance (E112) the patient has to pay the care in cash and then he/she can try to ask the insurance company to refund it, but there is no claim on the refund (article 20 Regulation of EU no. 883/2004).

The Czech Republic has also some contract about health care providing with certain states without EU, but mainly only urgent and pressing care is there covered. Any other care has to be approved by the insurance company before providing.

For other cases, mainly for the cases when the no covered care is requested there is so called "exceptional" payment for in public health care. It is said that exceptionally also no covered care can be paid for if the care is the only possibility from the point of state of the health of the insured person. This care needed previous acceptance of the insurance company, only if there is no risk delay.

Conclusion

The payment for the health care from the Czech public insurance system is legislatively treated. In ambulant form it is fully on the consideration of the insured person, concerned the planned hospital care it is very recommended to apply of the previous acceptance the insurance company which after guarantees the payment.

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Psychosocial effects of cavernom V. Portae

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I. Introduction

Psychological and social consequences of prehepatic portal hypertension with cavernous transformation of the portal vein in children. This is a relatively rare disease typically associated with frequent and serious complications. It is important to realize that not only is the child as a patient affected by the psychological and social consequences of the disease, but also his/her entire family. Although this disease is often congenital or acquired in early childhood, it is usually diagnosed relatively suddenly in a patient with no previous episodes. The disease is often diagnosed due to serious complications that directly threaten the life of the child (such as bleeding from the oesophageal varices or symptoms of an oncological disease). This fact means that the family has no time to prepare for the new situation and coping with such a shock may take quite a long time. This paper covers two case studies. Namely a boy who was diagnosed with prehepatic portal hypertension with cavernous transformation of the portal vein at the age of six based on bleeding from the oesophageal varices. The other case study concerns a seven-year-old girl whose diagnosis was determined based on extreme splenomegaly and thrombocytopenia at the age of five.

II. Case Study No. 1

II.I. The health condition of the boy

The boy is now 9 years old and had never been seriously ill. When he was six, one evening he did not feel well and had an episode of hematemesis. He was transferred from the hospital in České Budějovice to Thomayer Hospital in Prague. He was put in a medically-induced coma and the oesophageal varices were treated. During this haemorrhagic stroke lasting 14 days the boy bled a total of four times, always at attempted extubation. It was not possible to stop the bleeding endoscopically and for this reason the boy was transported to the Teaching Hospital in Hradec Králové where he had a transjugular intrahepatic portosystemic shunt (TIPS) implanted. He was then awoken from the medically-induced coma and during the following three weeks he vomited partially digested blood approx. 60 times a day. Physicians were preparing the boy's parents for the possibility that he would not survive the transport and operation. The boy remembers the time after awakening from the medically-induced coma, cannula cleaning, etc. Now, after the re-implantation of the TIPS, the values of liver tests fluctuate as the liver is not able to break down harmful substances. Owing to his impaired immunity, the boy is suffering from frequent infections threatening the TIPS. For this reason, it is necessary to administer prophylactic treatment based on broad-spectrum antibiotics in the case of any infection. This results in an increased quantity of toxic substances that the impaired liver fails to break down. Such a situation causes nausea and other subjective as well as objective difficulties. At the present time a cardiac murmur is appearing which is believed to have a connection with the TIPS.

II.II. The psychological and social state of the boy and his family

After the haemorrhagic stroke, the boy started suffering from considerable developmental regression. The boy had to learn to speak and walk again. At the beginning the boy was terrified of the physicians. With the passage of time he has got his fear under control and now he only tries to "negotiate" instead.

The boy experienced the progression of the disease at the beginning of school attendance when relationships between schoolmates are built. This situation resulted in exclusion from the school team and social isolation of the child. Moreover, when the boy's state of health allowed his return to school, his parents had to fulfil various conditions necessary for his return, such as a protective stomach belt, fender, etc. imposed by the school. The parents refused to fulfil such requirements arising from fears for the boy's safety and the boy started to attend school. Nevertheless, recurring absence due to appointments with the physician and recurring infections due to impaired immunity keep the boy out of the team and cause his social isolation. It is also important to realize that the physiological independence of the child is prevented by his health troubles and high risk of bleeding complications in the case of injury. The consequence is a strong parent fixation and frustration on the part of the boy. It is not possible for him to pursue activities suitable for his age, such as skateboarding. The aforementioned facts cause long-term frustration which has been transforming into deprivation that induces changes in mood towards a depressive state when he bursts into tears.

The boy has an older sister who was at preadolescent age when the symptoms of her brother's disease emerged. The situation accelerated her maturing. A greater portion of time and care was spent on her ill brother, which must have been very difficult for her. The girl's mother says that her brother's disease stole her childhood. Now she partly substitutes "the adults". Considering the unpredictability of the disease and special needs concerning nutrition and defecation, the boy requires constant care. Especially during visits of the boy's biological father (the boy has been living with his stepfather since he was one year old), his sister substitutes for his mother and supervises the regime measures which are necessary for the stabilization of her brother's state.

The boy's parents are self-employed, which allows them to organize work depending on the current situation. However, this does not mean that they can afford not to work. The system is such that when the boy sleeps or is with his biological father, the boy's parents work. This induces their long-term sleep deficit. The thought that their situation is not fair comes to their minds very often: "This is one of the relatively frequent thoughts. That sometimes life is not fair. That such a small child just does not deserve it." These feelings keep accompanying the parents from when they learned of the diagnosis and the related consequences.

Considering the sudden progression of the until then latent disease the parents were exposed to extreme stress when their child suddenly started bleeding from the digestive tract. In the hospital in the place of their residence, the nurse on duty made them wait for about 30 minutes when the boy had already had his muscle tone decreased due to loss of blood. After transport to Thomayer Hospital and repeated unsuccessful attempts to stop the bleeding, the parents were warned that it was likely that their son would die. They were offered transport to the Teaching Hospital in Hradec Králové and were informed that the boy may not survive it. The stress reaction of the boy's mother caused her to be able to fully function in the given situation. As she said, she had no time to break down. The situation was more difficult for the boy's father who had to take care of the boy's older sister and "was given the opportunity" to cry and feel sad. Since then the family is socially isolated from time to time. The boy and his parents continue to have problems within the framework of social communication when they have to cope with totally different problems from those of the people around them. They are sometimes afraid of falling asleep and the boy panics when he sees a physician. The parents live in fear of their son's death whereas the boy has similar feelings only occasionally. The need of safety and certainty of life is not sufficiently saturated. This deficit is noticeable in particular in the boy's parents. The boy experiences occasional episodes of reactive aggression. The parents feel somewhat isolated.

The family is "secured" by a social welfare allowance, a so-called care allowance. Pursuant to the Social Service Act, persons who take care of other persons, who suffer from long-lasting adverse health issues and need the help of another individual to be able to ensure essentials of life, are eligible for the care

allowance. The extent of essentials of life is given by the act and the degree of dependence. The boy has been awarded second degree of dependence, i.e. medium dependence. This means that according to the opinion of the competent medical assessor, the boy is not capable of four out of five listed essentials of life (mobility, orientation, communication, nurture, getting dressed and putting on shoes, body hygiene, defecation, care for health, personal activities, housekeeping). Nevertheless, the situation may radically change in April 2018 when the boy will have to be "re-assessed" whether or not he is eligible for this allowance. Holding of the "ZTP" identity card issued to handicapped persons allowing them to park in reserved places, which is essential when visiting hospital where the boy goes once every three months for control examinations, depends on such re-assessment.

III. Case Study No. 2

III.I. The health condition of the girl

The girl is now 7 years old and had never been seriously ill. At the age of two she was operated on for an inguinal hernia with ovarian adhesions. Her psychomotor development at first with quick acceleration, became uneven consequently. At the age of four, while on holiday, she went through an intestinal infection when the physician diagnosed moderate splenic enlargement which was attributed to the infection. Subsequently, during a prophylactic check-up after five years, it was discovered that the splenic enlargement persisted. For this reason, the girl was sent to an ultrasonic examination. The examination confirmed an enlarged spleen. The liver was of normal size and both the liver and spleen were without any structural changes. One month later, the girl underwent a neurological examination upon a request filed by the nursery school that the girl attended. The neurologist stated no neurological findings, but nevertheless sent the girl to the Paediatric Clinic in Karlov. After the girl was examined there, it was ascertained that the spleen was enlarged by approx. 13 cm and the blood count suggested significant thrombocytopenia, moderate coagulopathy and an increased count of reticulocytes. The girl was transported to the Oncological Department of Motol University Hospital for the determination of a differential diagnosis for acute leukaemia and storage disorders. Malignity was excluded by bone marrow tapping. The girl underwent additional examinations and at the end of September of the same year the diagnosis was determined. The girl suffered from dyspeptic disorders, frequent vomiting, her thrombocytopenia was aggravated, and cholestasis started to appear. A diffuse lesion was found on the liver along with one oesophageal varix, approx. 4 cm long. The girl was operated on and had a Meso-Rex bypass, which was followed by a decrease in the size of the spleen and stabilization of the thrombocyte count. The dyspeptic disorders nearly disappeared.

III.II. The psychological and social state of the girl and her family.

When the girl was treated at the Haemato-Oncological Department, her developmental regression appeared in the area of defecation, and in playing and drawing skills. Before hospitalization regression was already found in her drawing skills (see pictures 1 to 3) where fully structured pictures degraded to so-called "octopus level" after hospitalization, i.e. return to contentless scribbling. Moreover, since approx. the age of four, a developmental speech disorder (dislalia) appeared, and based on an ECG examination, an isolated focus was ascertained in the speech centre region. The retrograde trends both in speech and graphomotor and drawing skills worsened after hospitalization. After intervention, incontinence stabilized. Nevertheless, regression was also obvious in the social behaviour of the child. Due to the size of her spleen, the girl had to discontinue her leisure time activities that she had been used to pursuing, such as horse riding, riding on her scooter, trampolining, etc. The girl responded to her state by reactive aggression, on a verbal level and also towards objects, such as kicking stones, etc... She must have felt socially isolated to a great extent and became afraid of death as something ultimate. The girl invented imaginary friends and set her hopes on Christianity when Jesus Christ became a friend of hers. She built up the idea that after she died she would bring presents to children with baby Jesus.

She hated physical examinations and at first she screamed and fought. When she was hospitalized in the Oncological and Haematological Department of Motol University Hospital the situation changed. The girl needed to know exactly what would be done and when. She needed to feel that she had the situation under control.

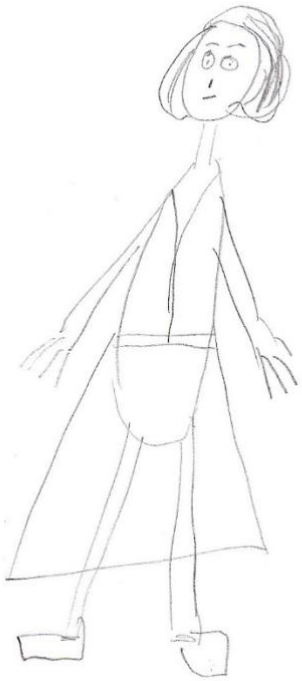
Her developmental regression ceased, and her state has been normalizing since the MR bypass operation. After she regained lucid consciousness, her speech abilities improved and after about two days after the operation she was able to produce structured drawings (see picture 4). The girl has symptoms of post-traumatic stress disorder.

The girl has four other siblings; three of the children are older and one sister is younger and was born after the girl's diagnosis was determined. The older siblings did not fully understand the girl's situation. Nevertheless, all the children had to become more independent consequently. For the younger sister it was difficult to keep her daily routine due to the repeated examinations and hospitalizations. The girl became fixed to her mother, which complicated the situation to a great extent.

Both parents have flexible working hours, which allowed them to accompany the girl within the framework of medical intervention without one of the parents having to give up their profession, which would result in a loss of their income for the family. In addition, the family lives in a two-generation house with the mother's parents, who assumed care for the older children.
Both parents say that at the moment when they were informed of a possible malignant disease, their life changed all of a sudden. The mother says that during the first two days in the Oncological Department she acted as if on "autopilot". She felt that it was like a dream and the whole situation seemed unrealistic to her. It reminded her of a surrealist painting. She experienced the same state once again after her daughter was moved to the Intensive Care Unit. For the father the most difficult situation was after the operation when the girl was no longer administered opiates and developed a fixed idea that she had dirty hand which she started to bite. The state lasted for about two hours.

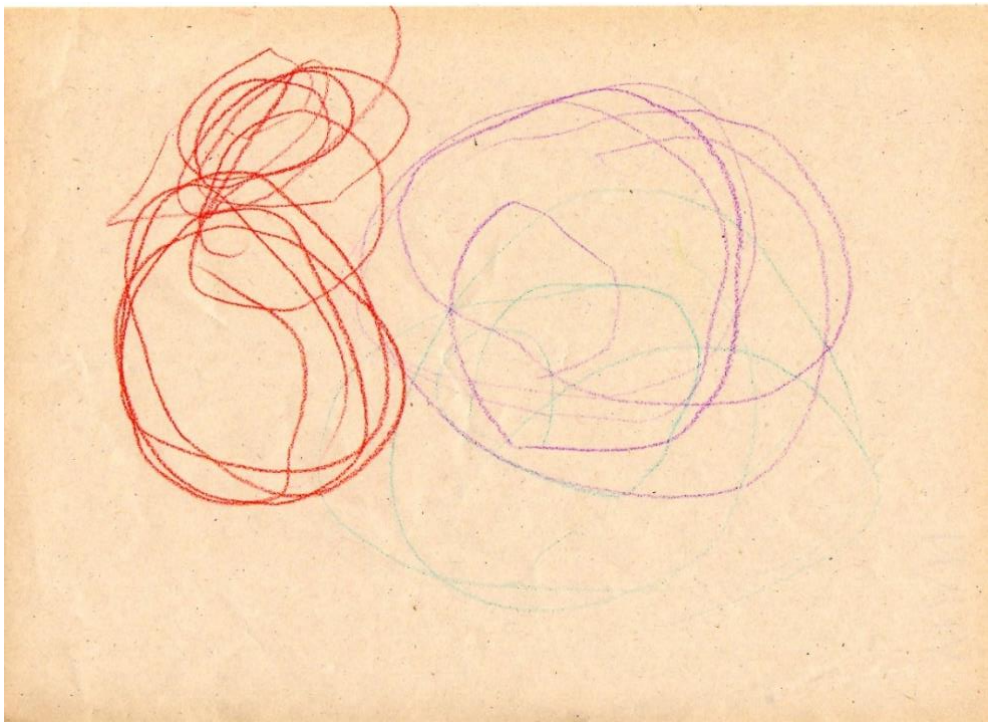
IV. Conclusion

Prehepatic portal hypertension with cavernous transformation of the portal vein causes multiple complications both at the medical as well as social and mental level. Repeated hospitalizations and the contingent character of the disease result in an insufficient saturation of vital needs such as safety, certainties of life and the need of social activity or positive open perspectives. The consequence is aggravated frustration which gradually transforms into deprivation. The consequence is developmental deviations, reactive aggression, and social isolation, which may little by little end in so-called social death. Prof. PhDr. Rudolf Kohoutek, CSc. has defined social death as follows: "chronic diseases, isolation from close relatives, loneliness, social deprivation (strain) often likely accelerating physical death". It is important to give a thought to the quality of life and perspectives of children who are provided symptomatic treatment. Such children will have to strictly adhere to regime measures. Girls are not likely to have children and if they decide to have them, they will risk that the increasing abdominal pressure will cause a bleeding attack that will threaten not only their own life but also the life of their child. Also work perspectives are difficult to foresee. The lives of the families whose child has this diagnosis is predominantly about medical reports, a suitcase ready to be taken to hospital and waiting when their child starts bleeding or suffers from failure of vital organs.



Picture No. 1

Picture No. 2



Picture No. 3

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