When is a child untransplantable?

Professor Nizam Mamode

Guys and St Thomas Hospital

Evelina Children's Hospital

Great Ormond Street Hospital

Figure 8.8 **Median waiting time to deceased donor transplant for paediatric patients registered on the kidney transplant list, 1 April 2011 - 31 March 2014**

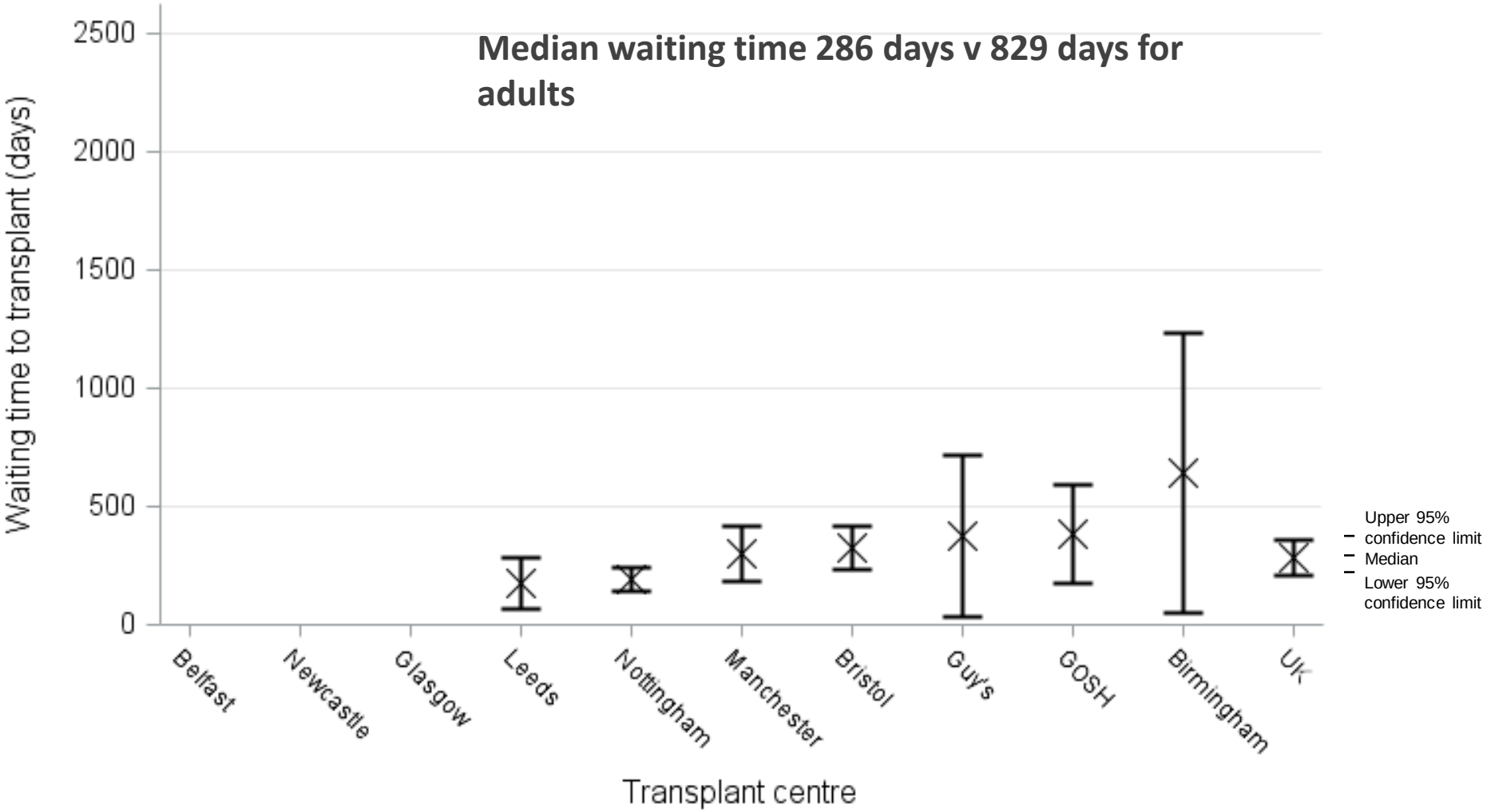
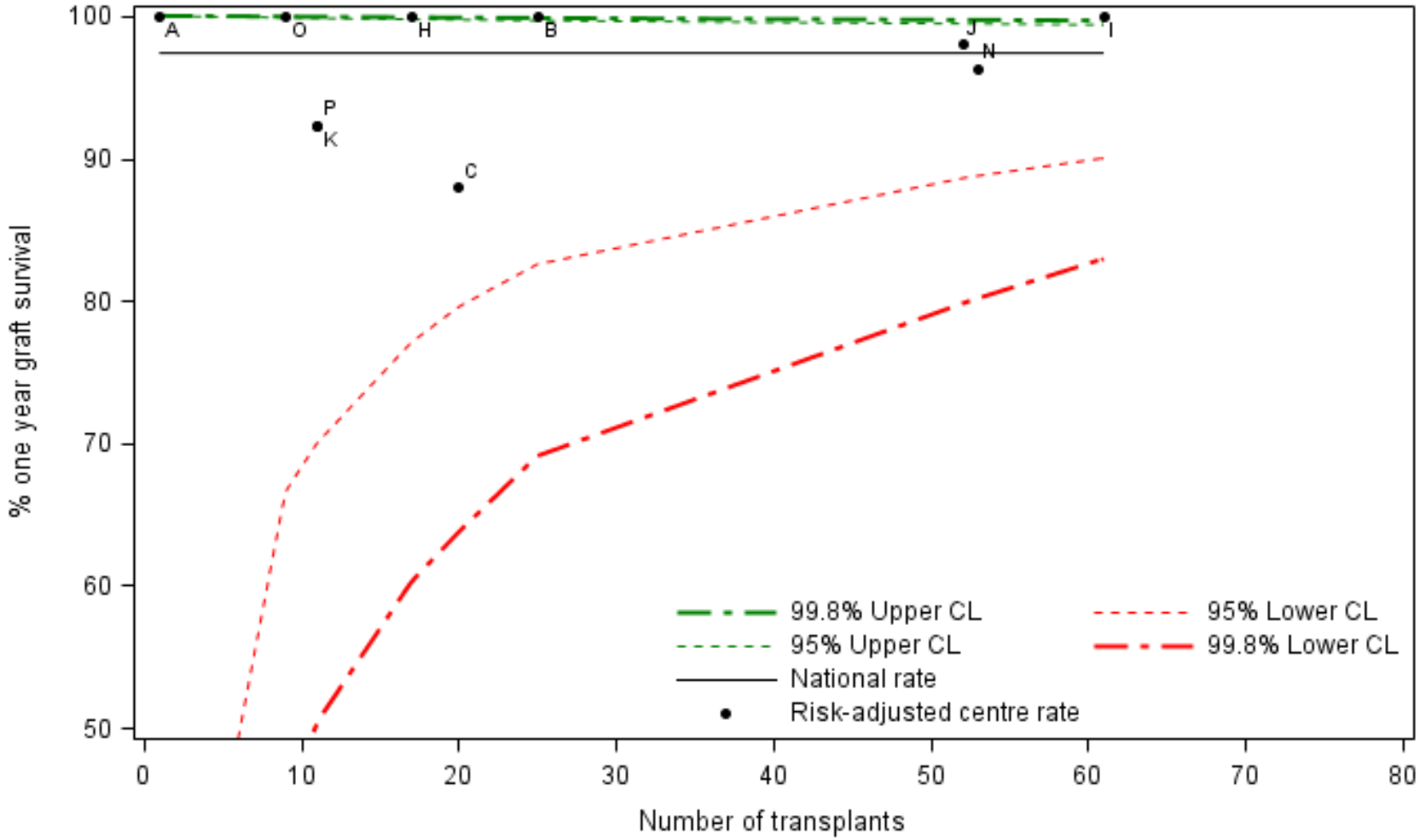


Figure 11.5 **Risk-adjusted one year graft (death censored) survival rates for first live donor kidney transplants in paediatric patients, between 1 April 2012 and 31 March 2016**

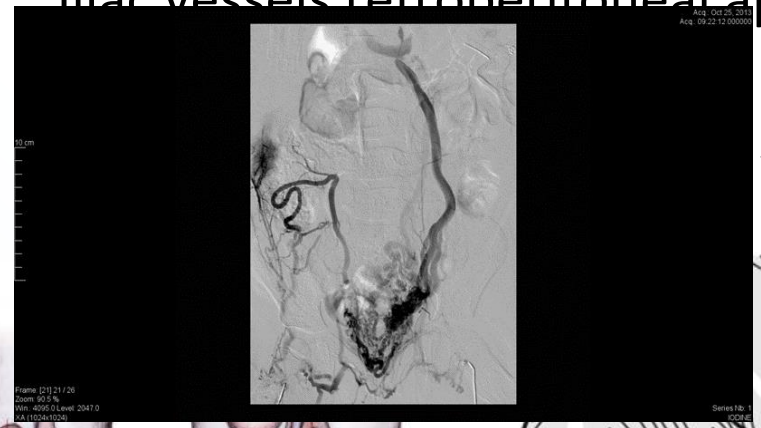


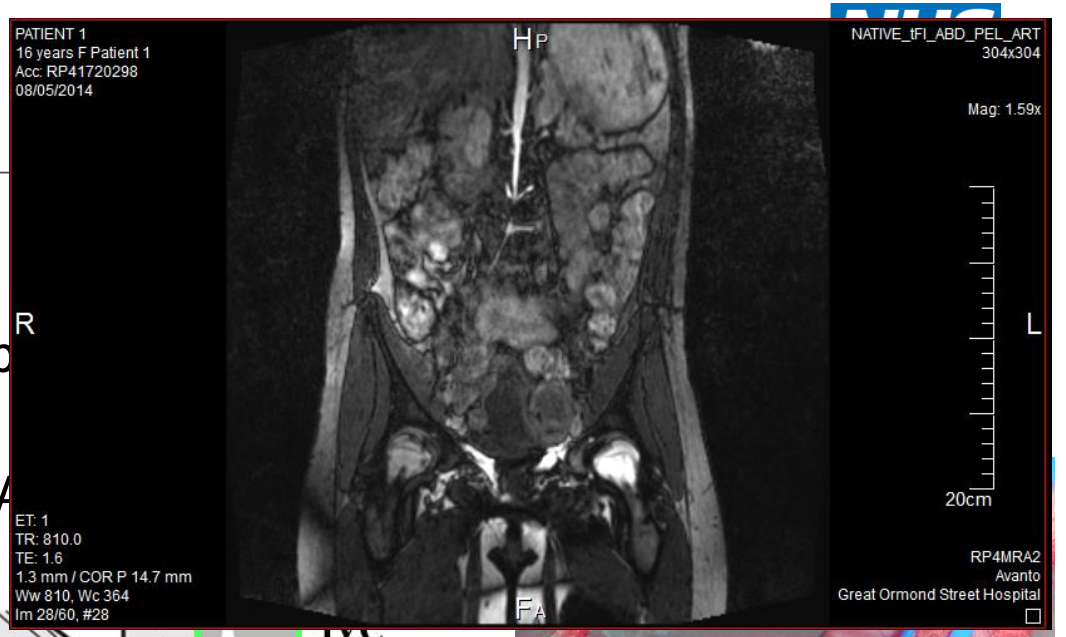
Source: Annual Report on Kidney Transplantation 2016/17, NHS Blood and Transplant

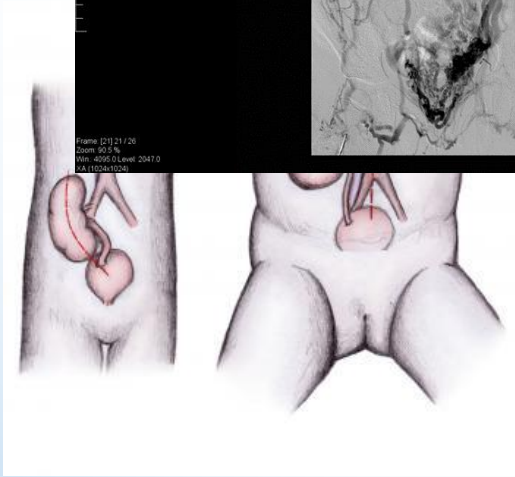
Implantation

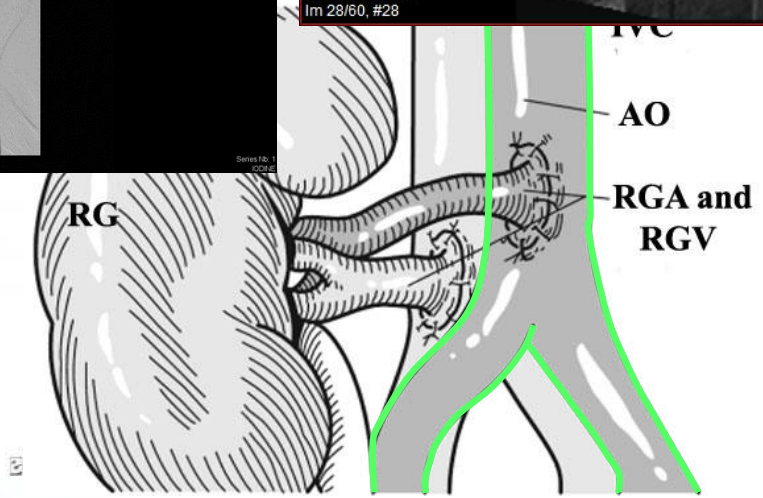
Children >20kg:

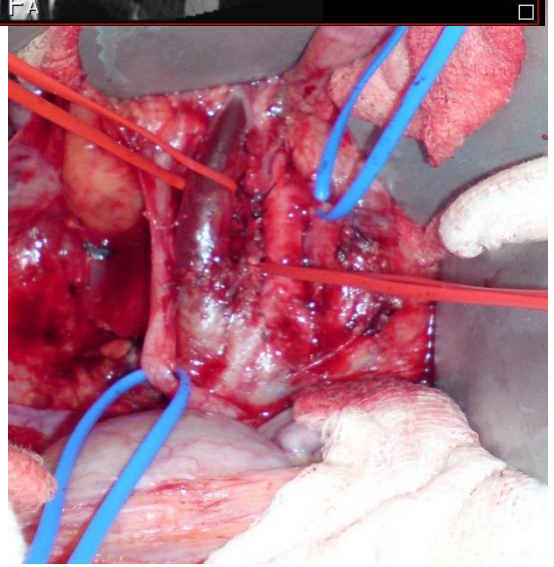
Iliac vessels retroperitoneal ap











Why do children have vascular anomalies?

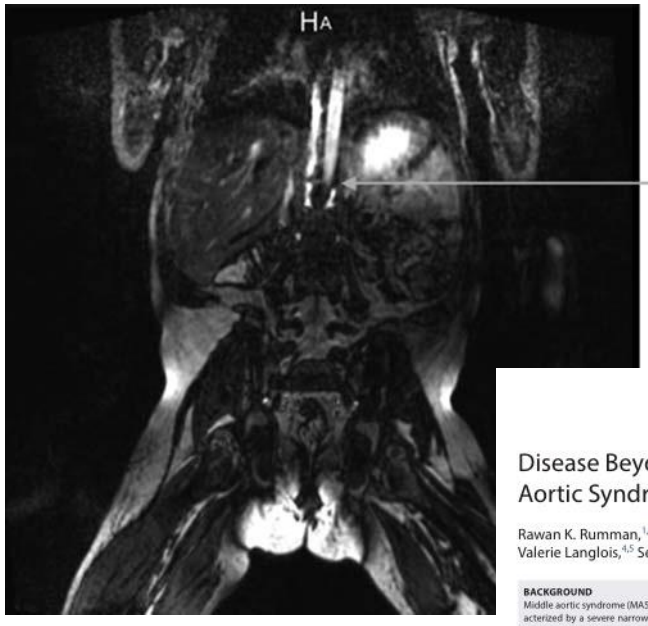
Central lines

Previous surgery

Hypercoaguability

Congenital disease:

Mid-aortic syndrome



STATE OF THE ART

Disease Beyond the Arch: A Systematic Review of Middle Aortic Syndrome in Childhood

Rawan K. Rumman,[1,2] Cheri Nickel,[3] Mina Matsuda-Abedini,[4,5] Armando J. Lorenzo,[5,6] Valerie Langlois,[4,5] Seetha Radhakrishnan,[4,5] Joao Amaral,[5,7] Luc Mertens,[5,8] and Rulan S. Parekh,[4,5,9]

BACKGROUND
Middle aortic syndrome (MAS) is a rare clinical entity in childhood, characterized by a severe narrowing of the distal thoracic and/or abdominal aorta, and associated with significant morbidity and mortality. MAS remains a relatively poorly defined disease. This paper systematically reviews the current knowledge on MAS with respect to etiology, clinical impact, and therapeutic options.

METHODS
A systematic search of 3 databases (Embase, MEDLINE, and Cochrane Central Register of Controlled Trials) yielded 1,252 abstracts that were screened based on eligibility criteria resulting in 184 full-text articles with 630 reported cases of childhood MAS. Data extracted included patient characteristics, clinical presentation, vascular phenotype, management, and outcomes.

RESULTS
Most cases of MAS are idiopathic (64%), 15% are associated with Mendelian disorders, and 17% are related to inflammatory diseases. Extra-aortic involvement including renal (70%), superior mesenteric (30%), and celiac (22%) arteries is common, especially among those with associated Mendelian disorders. Inferior mesenteric artery involvement is almost never reported. The majority of cases (72%) undergo endovascular or surgical management with residual hypertension reported in 34% of cases, requiring medication or reintervention. Clinical manifestations and extent of extra-aortic involvement are lacking.

CONCLUSIONS
MAS presents with significant involvement of visceral arteries with over two thirds of cases having renal artery stenosis, and one third with superior mesenteric artery stenosis. The extent of disease is worse among those with genetic and inflammatory conditions. Further studies are needed to better understand etiology, long-term effectiveness of treatment, and to determine the optimal management of this potentially devastating condition.

Keywords: abdominal aorta; blood pressure; bypass; coarctation; endovascular; grafting; hypertension; middle aortic syndrome; renal artery stenosis.

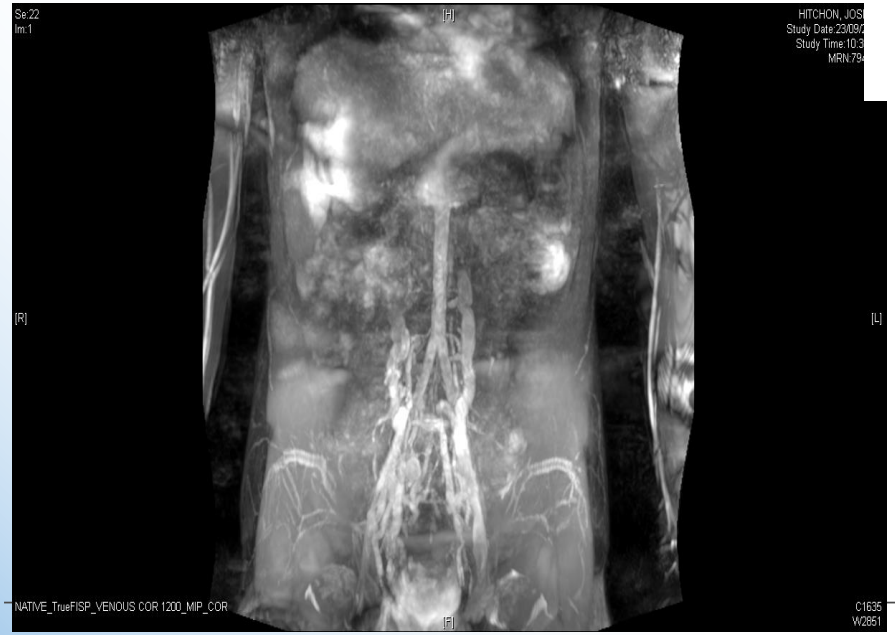
doi:10.1093/ajh/hpu296

Am J Hypertens. 2015Jul;28(7):833-46.

15yr old boy

Weight 55kg

ESRD due to pneumococcal HUS



Left LIF incision with EIV and EIA anastomosis Immediate function but slow venous outflow noted hence started on LMWH

Day 15 post Tx had a non occlusive thrombus in main renal vein at the hilum

6 weeks of dalteparin, resolved, renal function fine

Last eGFR 44ml/min/1.73m2

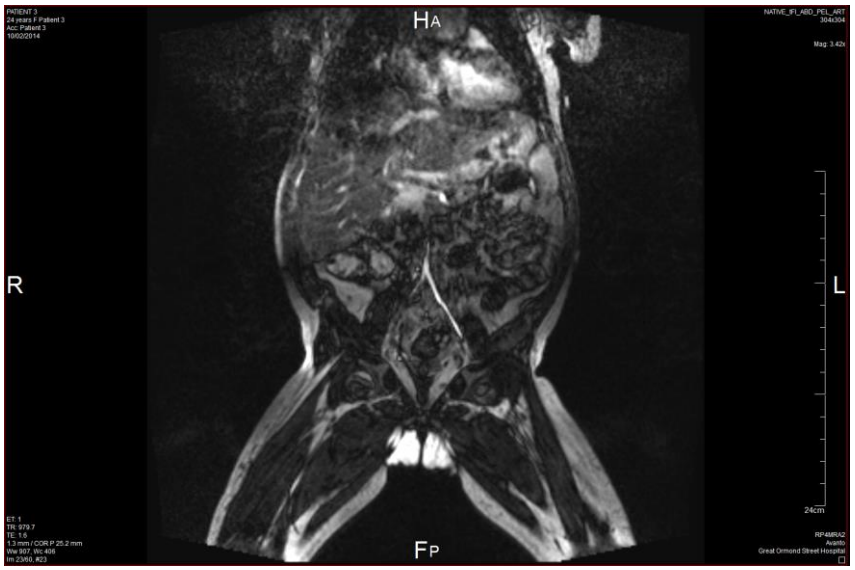
7 year old girl, congenital nephrotic syndrome

Bilat nephrectomies aged 2 years

LD transplant aged 3 years, failed: thrombosis

Recurrent line thromboses, SVC thrombosis

Grandmother wants to donate



Midline incision

Dense adhesions

Suprarenal IVC accessible

Transplanted onto aorta and IVC, with periop heparin

Options if no IVC

American Journal of Transplantation 2017; 17: 1670–1673
Wiley Periodicals Inc.

doi: 10.1111/ajt.14213

Case Report

Mobilise liver and use retrohepatic IVC/R hepatic vein

Mesenteric veins

Patel AJT 2003

Collateral veins

Successful Renal Transplantation in Small Children With a Completely Thrombosed Inferior Vena Cava

P. Verghese[1], E. Minja[2], V. Kirchner[2], B. Chavers[1], A. Matas[2] and S. Chinnakotla[2,*]

[1]Department of Pediatrics, University of Minnesota Medical School, Minneapolis, MN
[2]Department of Surgery, University of Minnesota Medical School, and University of Minnesota Masonic Children's Hospital, Minneapolis, MN
**Corresponding author: Srinath Chinnakotla, chinni@umn.edu*

technical difficulty of achieving renal venous outflow drainage and the risk of graft thrombosis. Without a transplant, these children are relegated to a probably shortened life on dialysis, with its associated problems (including the need for multiple dialysis-access procedures).

We report our experience with two small children who had completely thrombosed IVC. Using a new technique to anastomose the renal vein to the right hepatic vein/IVC junction, we implanted an adult-sized graft in each patient. We believe that long-term success is likely for

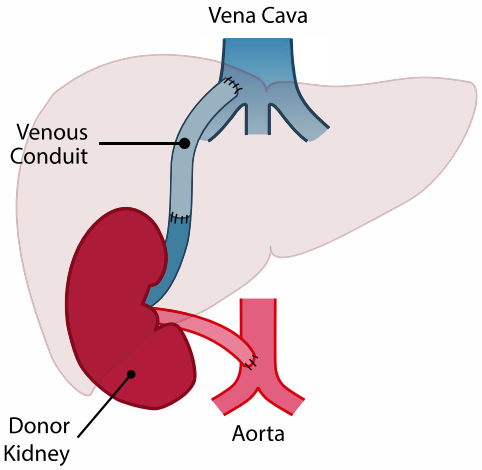


Figure 1: Diagram showing the site of the renal vein and renal artery anastomosis.

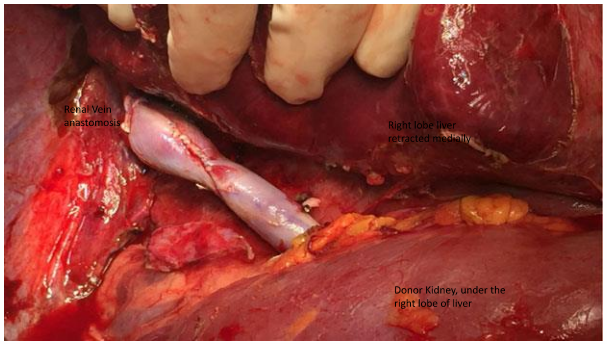


Figure 3: Intraoperative photograph showing the renal vein conduit anastomosed to the right hepatic vein/inferior vena cava junction and the final position of the kidney graft under the right lobe of the liver.

Portal vein

Cauley Pediatr Transplant 2013

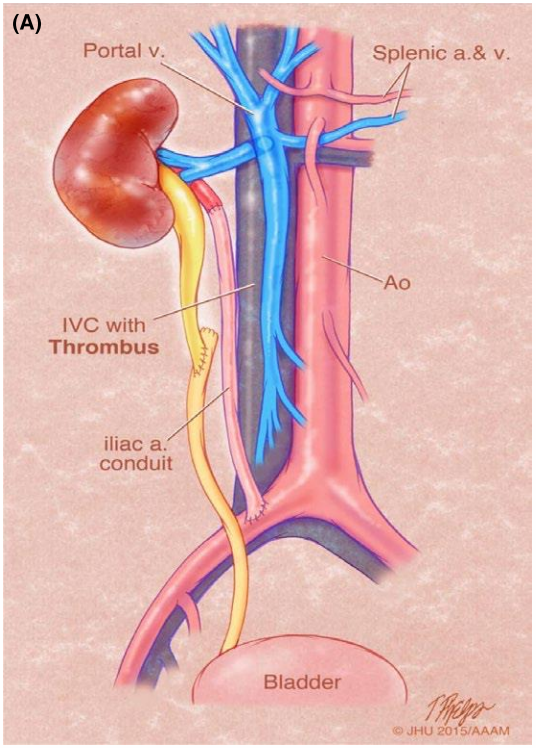
Accepted: 23 September 2017
DOI: 10.1111/ctr.13127

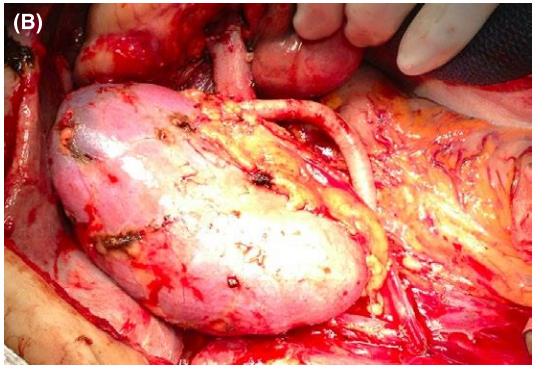
ORIGINAL ARTICLE

WILEY Clinical TRANSPLANTATION
The Journal of Clinical and Translational Research

Nontraditional sites for vascular anastomoses to enable kidney transplantation in patients with major systemic venous thromboses

Bonnie E. Lonze[1] | Nabil N. Dagher[1] | Nada Alachkar[2] | Annette M. Jackson[3] | Robert A. Montgomery[1]

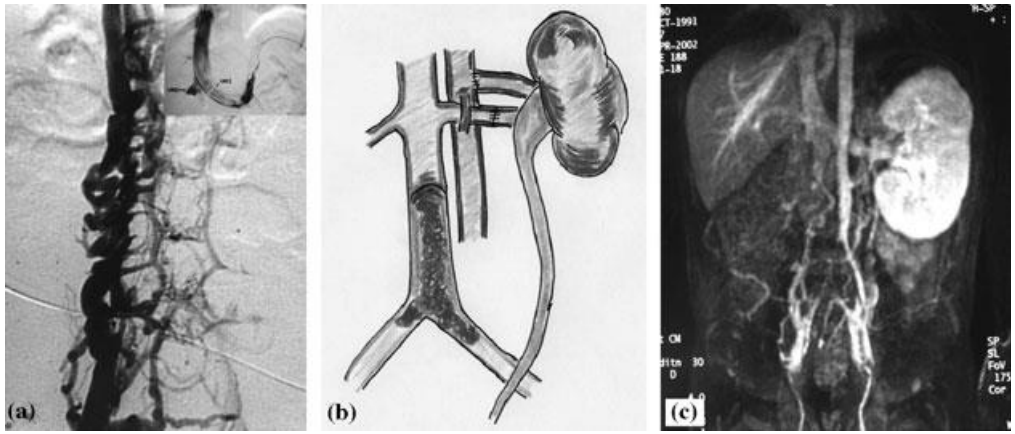




Orthotopic approach

Martinez-Urrutia Pediatr Transplant 2007

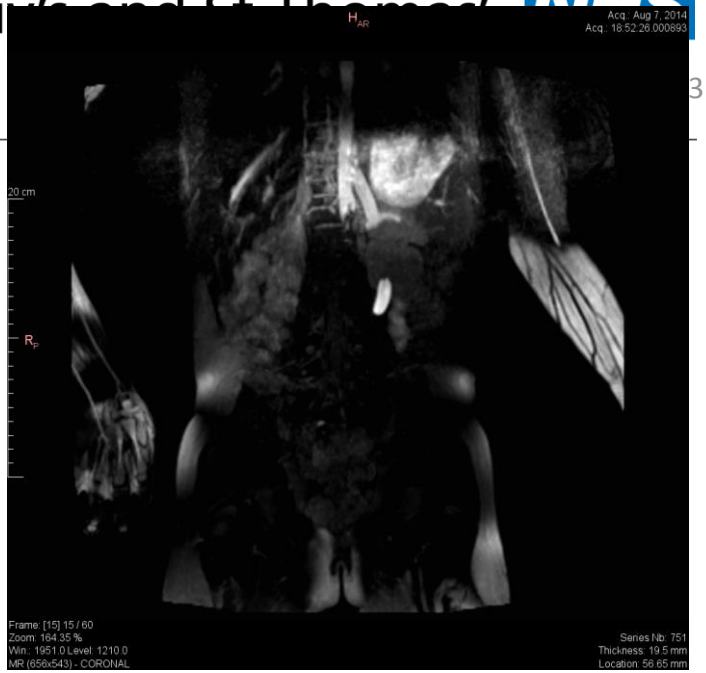
3 patients using adult kidneys



It may be worth exploration

Consider starting recipient prior to the donor

May benefit from 2 experienced recipient surgeons and 1 experienced donor surgeon

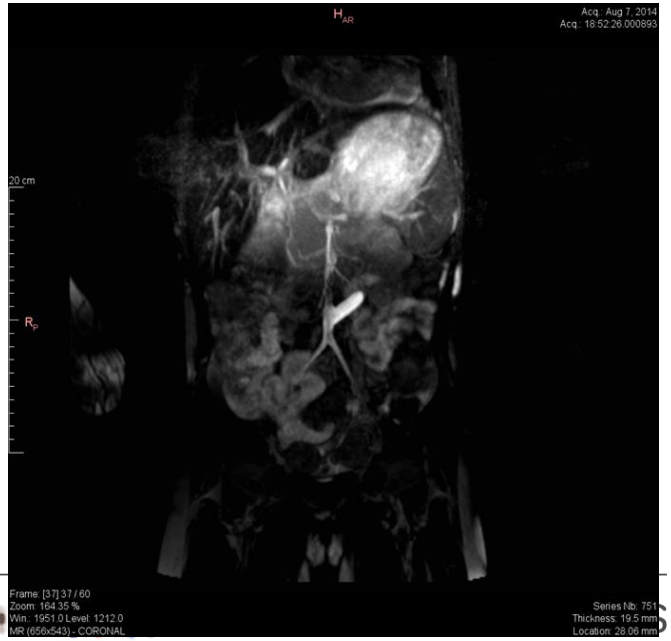


9 year old girl, 24kg

Previous right nephrectomy (dysplasia)

Mid-aortic syndrome

4 months previously had graft: supra-coeliac aorta to bifurcation, with L nephrectomy (thoraco-abdominal incision)



49 yr old father is donor, 000MM

Midline incision

Inspection of vessels

Transplant onto L CIA

17 year old girl, 34 kg

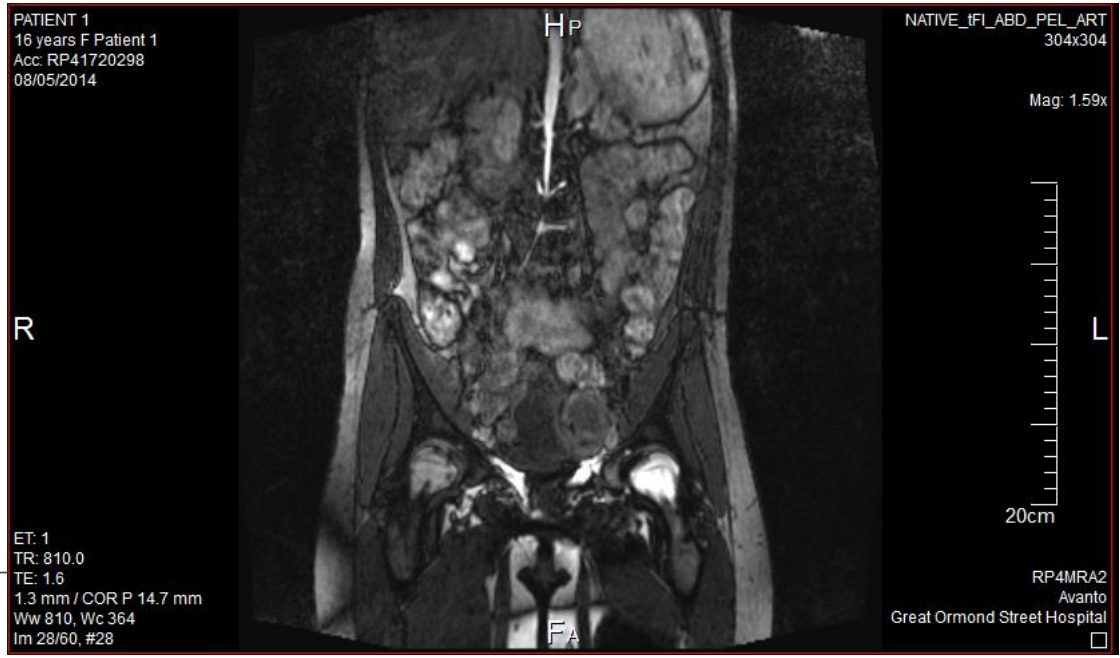
CAKUT, aortic coarctation, biltateral hydronephrosis, UTIs

At 2 years: graft to aortic arch (twice)

Ureters re-implanted, uretero-ureterostomy, augment, re-augment and Mitrofonoff (7 years)

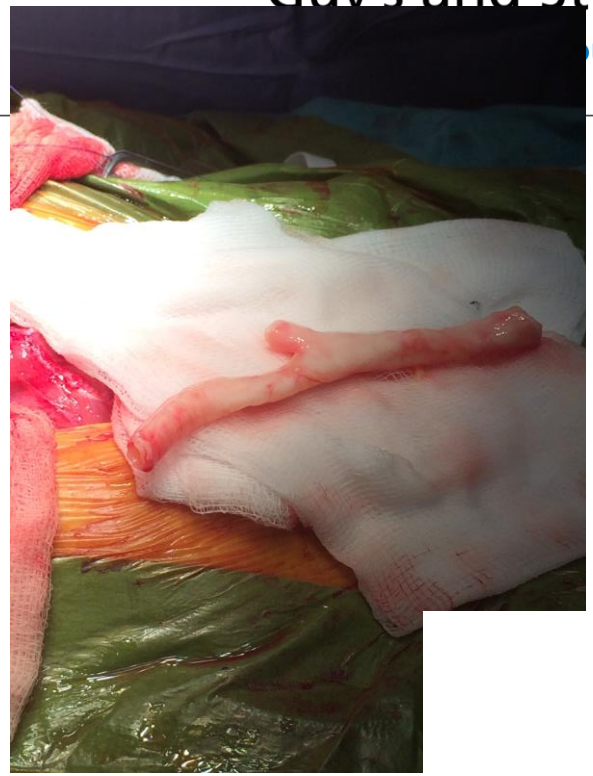
At 16 years: nephrectomy- HD

Mother is donor



Surgery

Recipient before donor



Dense adhesions

Split diaphragmatic crura

Deceased donor iliac artery conduit

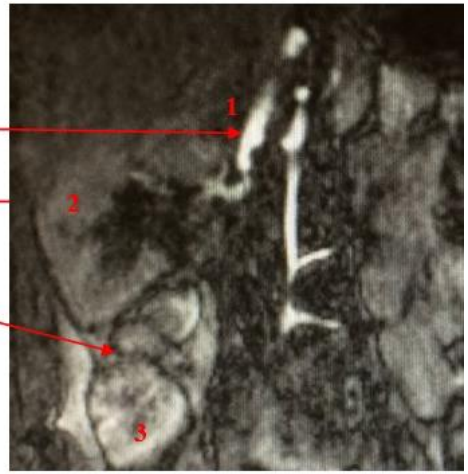
to supra-coeliac aorta,

tunneled under porta hepatis

Renal vein to IVC

Ureter to augmented bladder

Figure 5



Issues with DD conduit

Patient type -
A1,A2;B8,B44;Bw4,Bw6;Cw5,Cw7;DR8, DR11,DR52;DQ7

Kidney type -
A2,A30;B13,B44;Bw4;Cw5,Cw6;DR11,DR52;DQ7

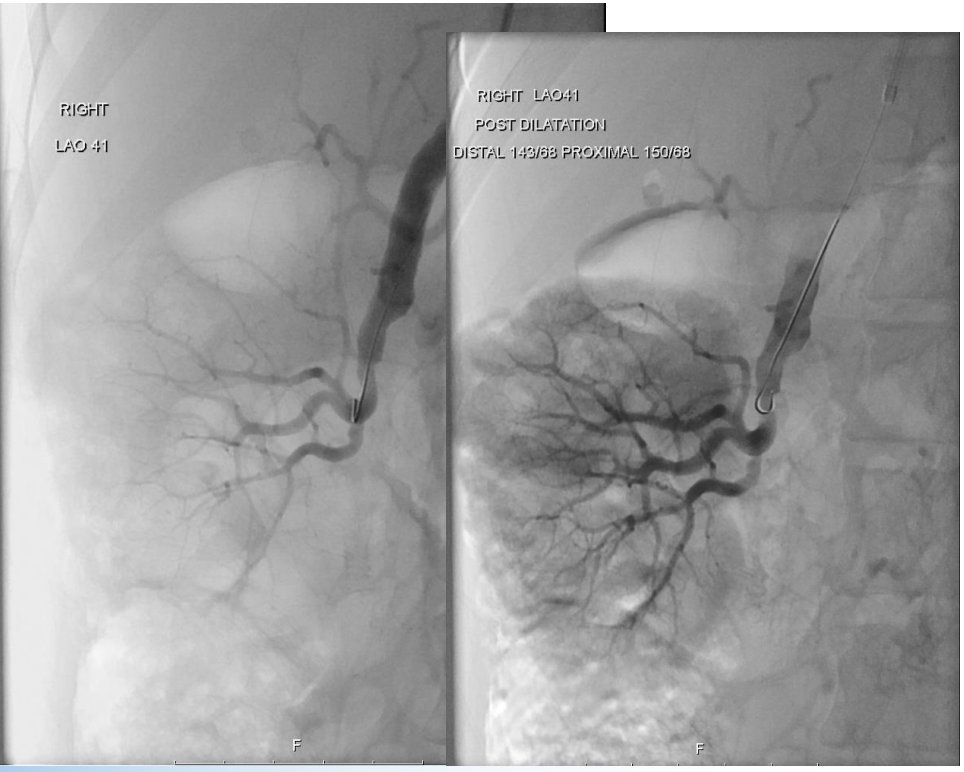
Iliac graft type – A1, A3;B35,57;Bw4,Bw6;Cw4,Cw6;DR1,DR7,DR53;DQ5,DQ9

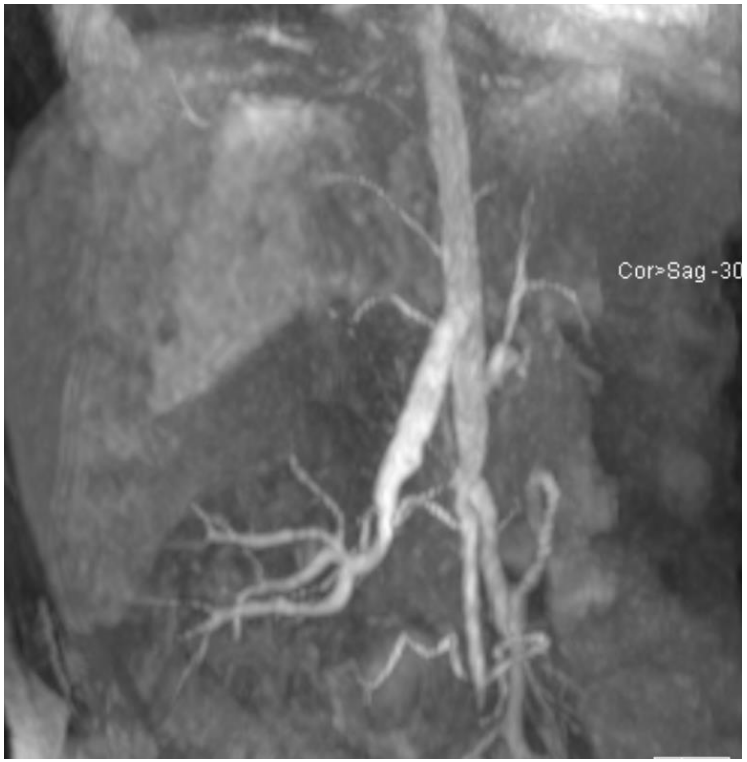
Developed rise in Cr associated with small rise in DSA to iliac conduit

No DSA to kidney

? Rejection of the conduit

Nov 2016- PTA for stenosis March 2017





6 year old girl 18kg

Cause of renal failure: 'renovascular'

Previous aortic graft

On haemo-dialysis (line)

SVC obstruction

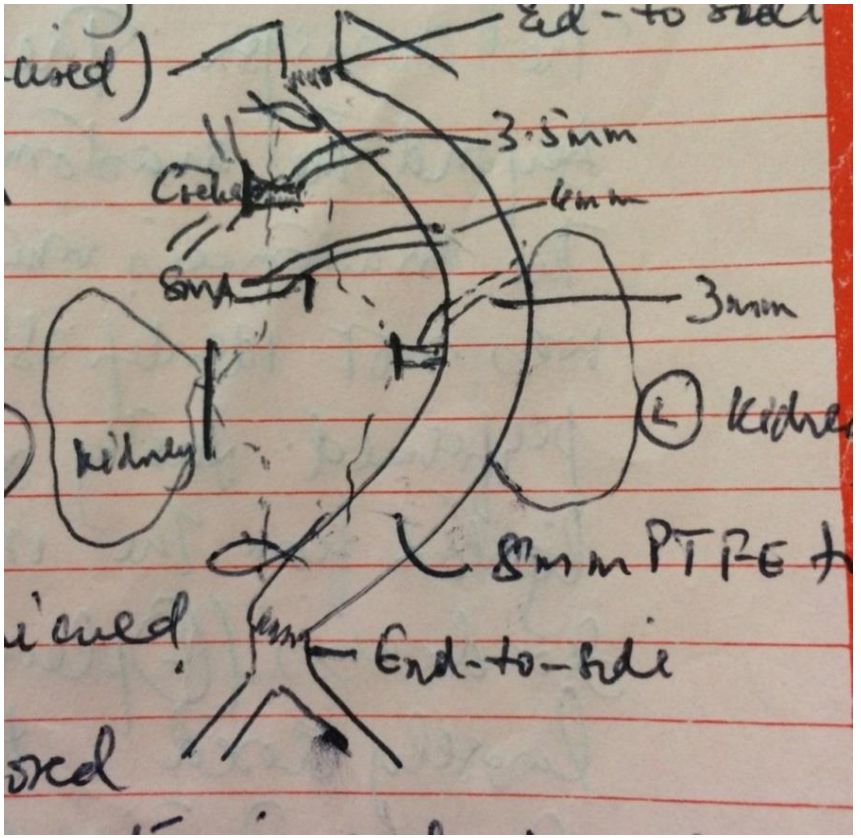
- considered unlikely any further access for dialysis available

Major abdominal trauma at age 4:

- multiple laparotomies (4)
- tracheal stenosis, CVA

Blood group incompatible donor:

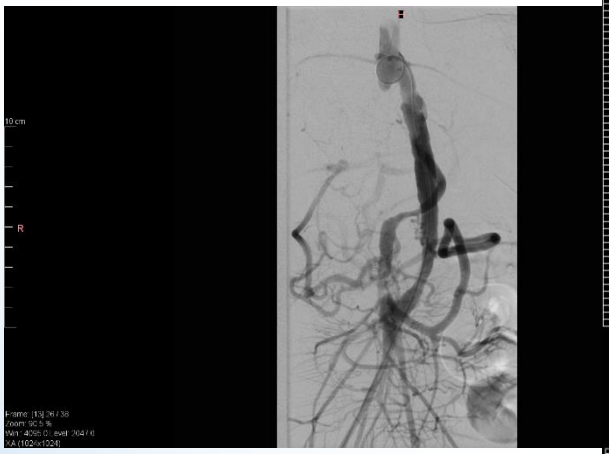
- mother A into O, titres 1in 8

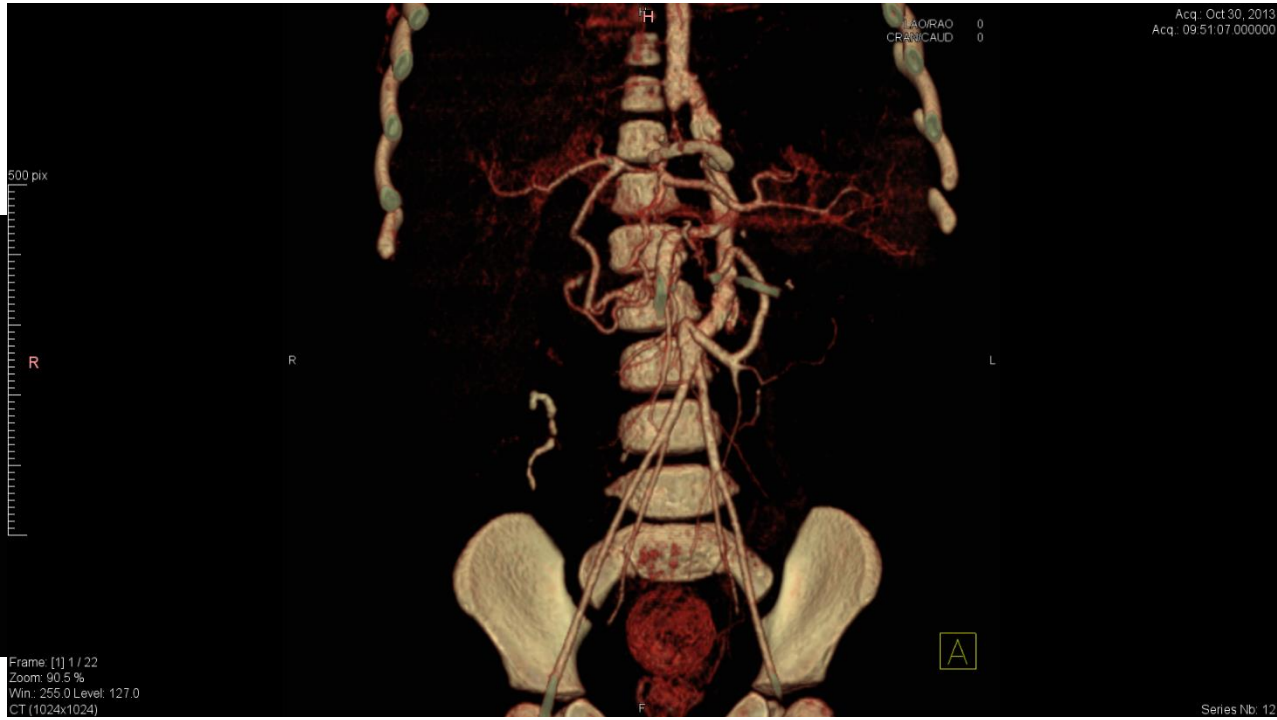


Highly sensitised CRF 87%

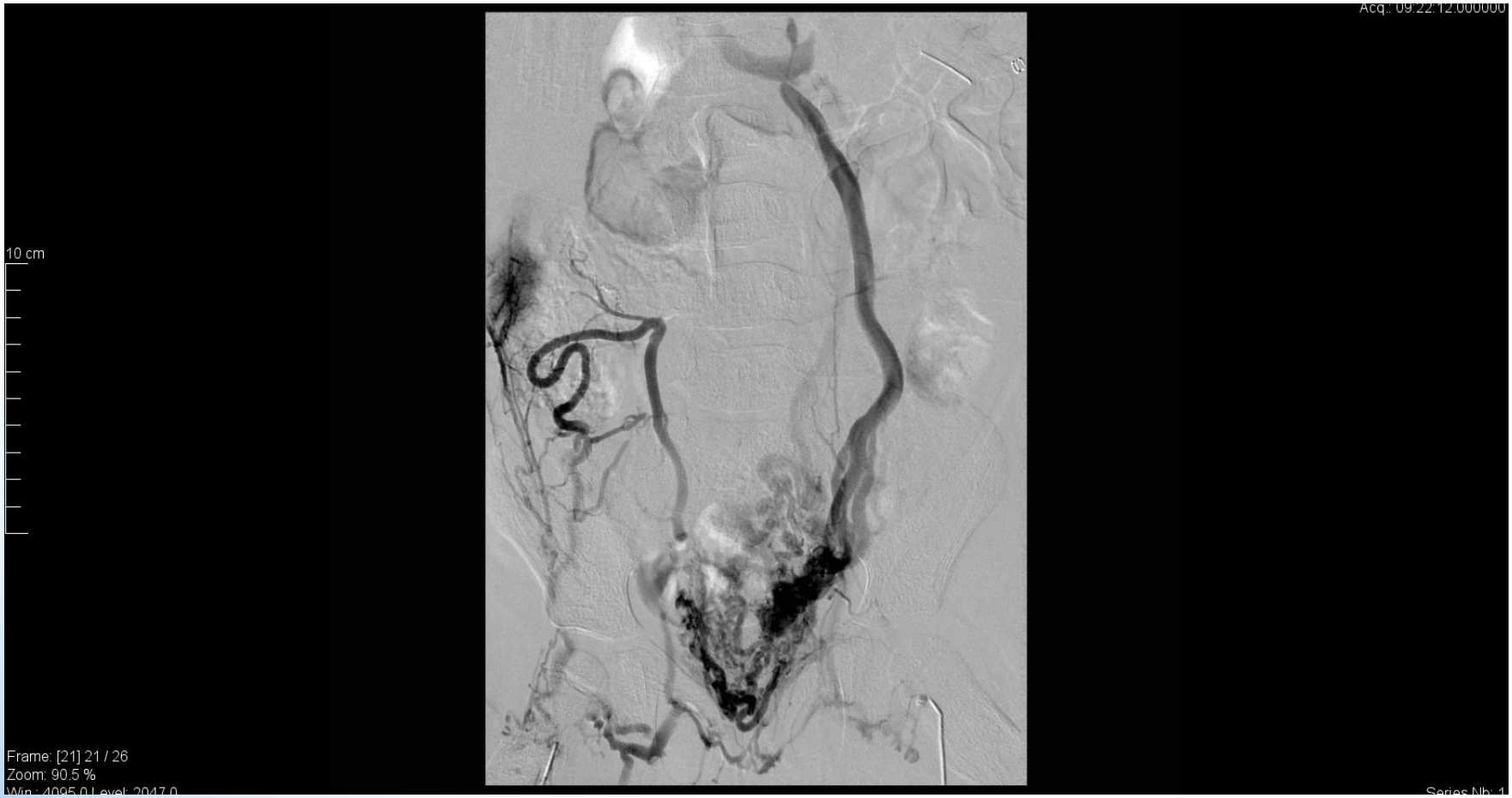
Investigations

Angiogram





Venous anatomy



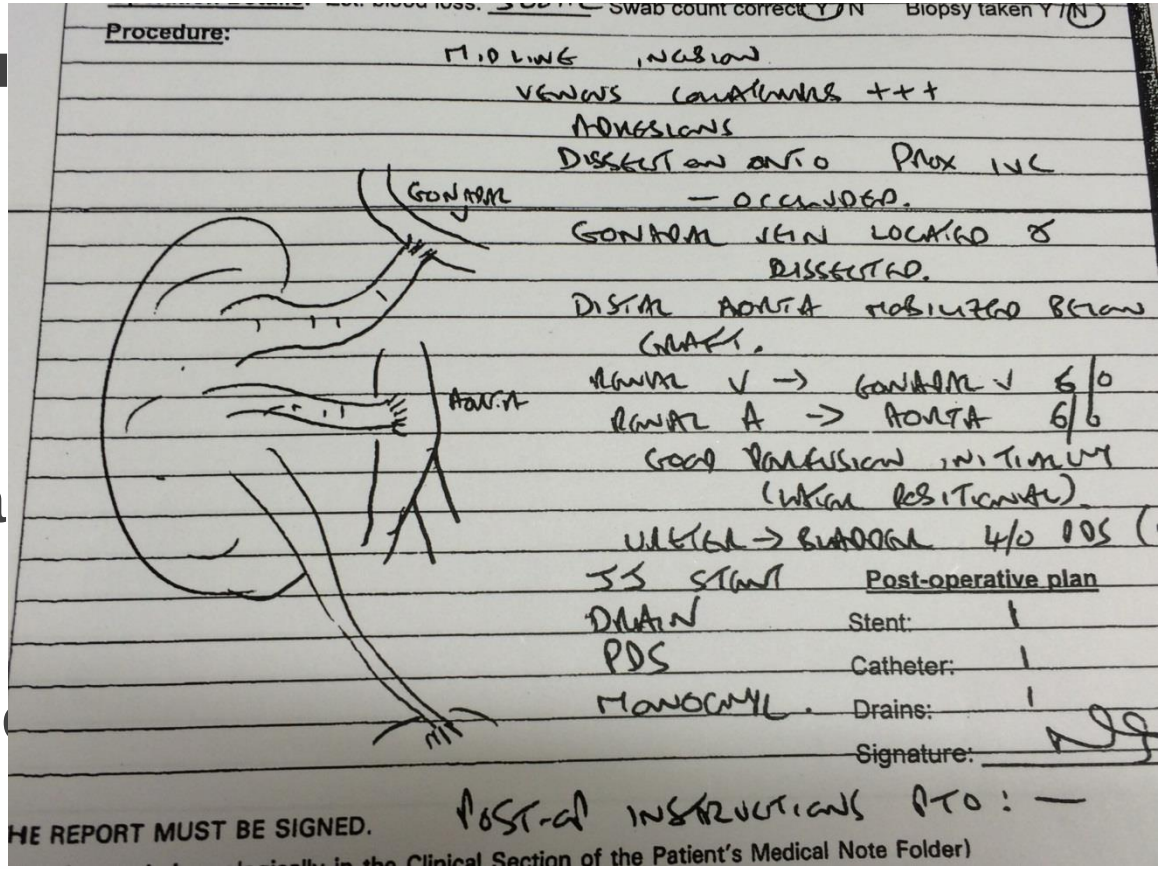
Our approach

Multiple MDMs at adu

Hospital ethics team

Palliative care team a

2 consultants at surg



Donor surgery started after recipient

Guy's and St Thomas' NHS
NHS Foundation Trust

Insights in Transplanting Complex Pediatric Renal Recipients With Vascular Anomalies

Pankaj Chandak, MRCSEng,[1] Nicos Kessaris, FRCS,[1] Chris J. Callaghan, FRCS,[1] Francis Calder, FRCS,[1] Jelena Stojanovic, MRCPCH,[1] Jonathon Olsburgh, FRCS,[1] Martin Drage, FRCS,[1] Helen Hume-Smith, FRCA,[2] Zubir Ahmed, MRCS,[1] Anna Adamusiak, MRCS,[1] Derek Roebuck, FRCR,[3] Colin Forman, FRCS,[4] Stephen D. Marks, FRCPCH,[5] and Nizam Mamode, FRCS[1]

Background. Children with end-stage kidney disease may have coexisting iatrogenic or congenital vascular anomalies making transplantation difficult. We describe our approach in 5 recipients with vascular anomalies and significant comorbidities, including one case of blood group incompatibility. **Methods.** Five children aged 3 to 17 years (median, 7 years), weighing 14 to 34 kg (median, 18 kg) kg of whom 4 had occluded inferior vena cava or iliac veins and 2 had previous complex vascular reconstructions before transplantation for midaortic syndrome and multiple aortic aneurysms, respectively underwent renal transplantation. To establish implant feasibility surgery was commenced in 2 recipients before the donor surgery. **Results.** There was 4 (80%) of 5 patient survival after 1 death from sepsis (with a functioning graft) and 2 cases of delayed graft function. At the latest median follow-up of 19 months, there was 100% (death-censored) renal allograft survival with estimated glomerular filtration rates (mL/min per 1.73 m^2) of 43 to 72 (median, 55). **Conclusions.** We conclude that major vascular anomalies do not necessarily preclude transplantation in complex pediatric patients and that surgical exploration of the recipient before commencing the donor surgery is valuable where feasibility and safety are uncertain. In addition, we have developed a novel classification system of congenital vascular abnormalities and propose its use in complex pediatric transplantation.

(*Transplantation* 2017;101: 2562–2570)

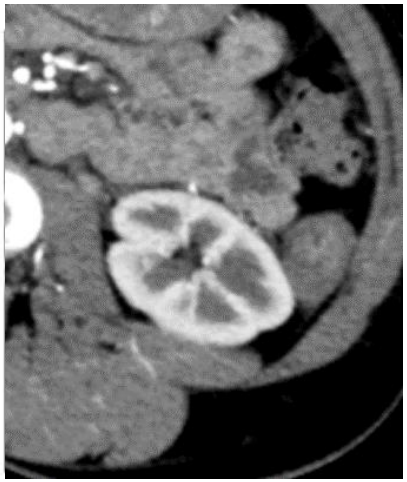
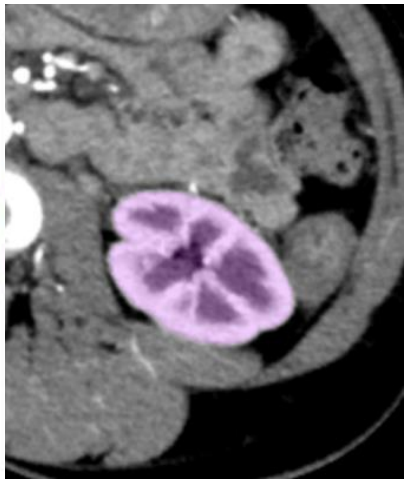
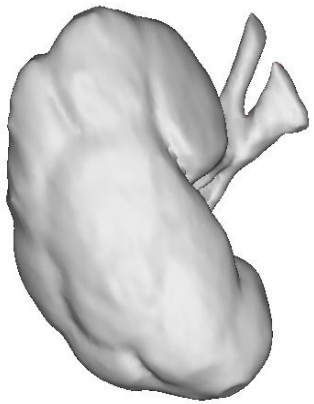
Abnormality present	Aorta (A)	IVC (V)
Entire abdominal vessel patent	A1	V1
Infrarenal segment occluded, absent or narrowed	A2	V2
Suprarenal segment occluded, absent or narrowed	A3	V3
Entire abdominal vessel occluded, absent or narrowed	A4	V4

Footnotes:
Aorta refers to abdominal aorta from diaphragm to aortic bifurcation
IVC refers to abdominal inferior vena cava from diaphragm to IVC bifurcation

KING'S HEALTH PARTNERS

"How can we make the surgery safer for complex kidney transplantation in children"?

Using 3D printing of paediatric abdominal structures and adult donor kidneys

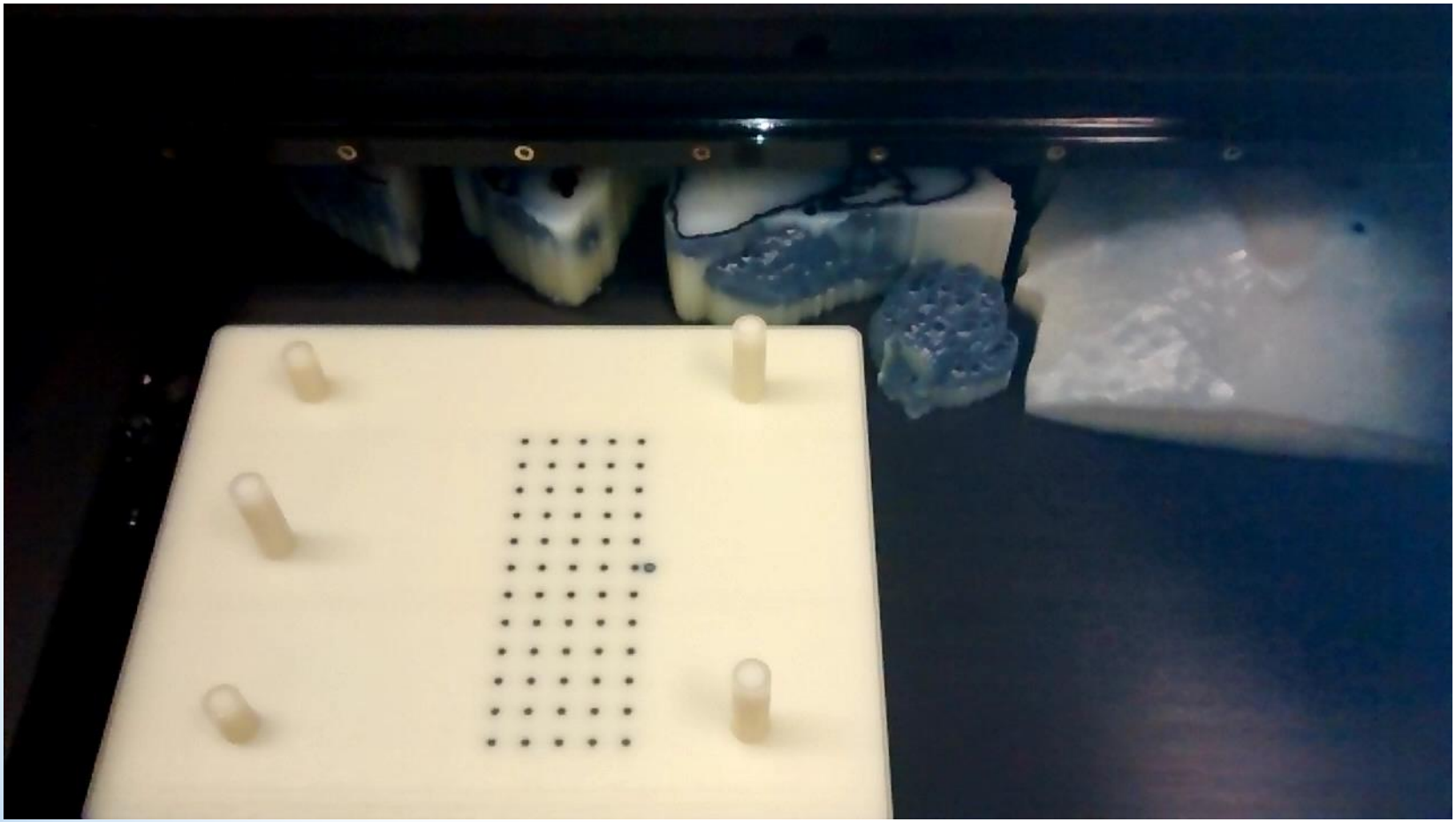
Image acquisition	Image segmentation	Computer aided design	Additive manufacture
			
3D CMR 3D CT	Thresholding Region growing Manual editing	Hollowing Cutting Labelling	3D printing Stereolithography Laser sintering

Software: Mimics Medical 18.0 (Materialise, 2015)

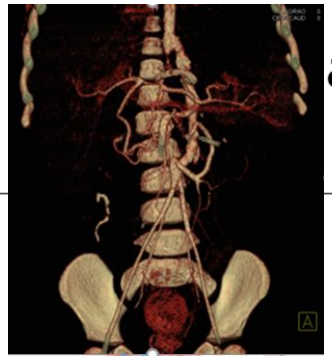
Flexible materials: TangoPlus FullCure930, Stratasys

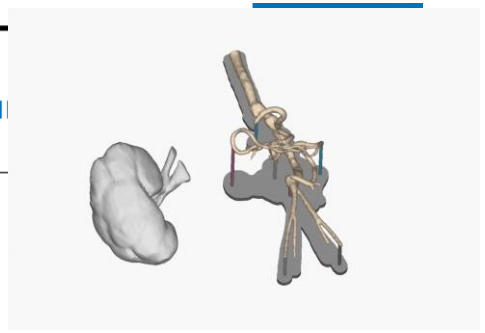
Rigid materials: VeroWhitePlus FullCure835, Stratasys

3D Printer: Stratasys Objet500 Connex1 printer

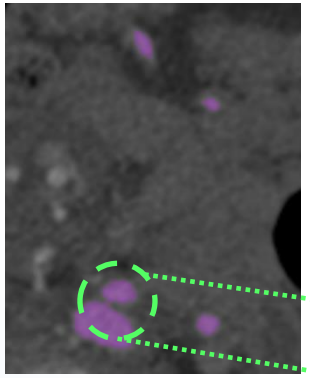


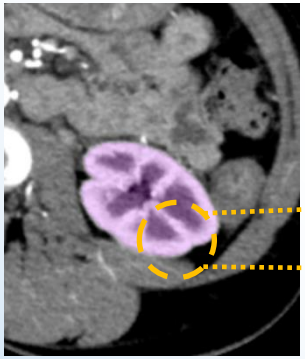
Proof of concept

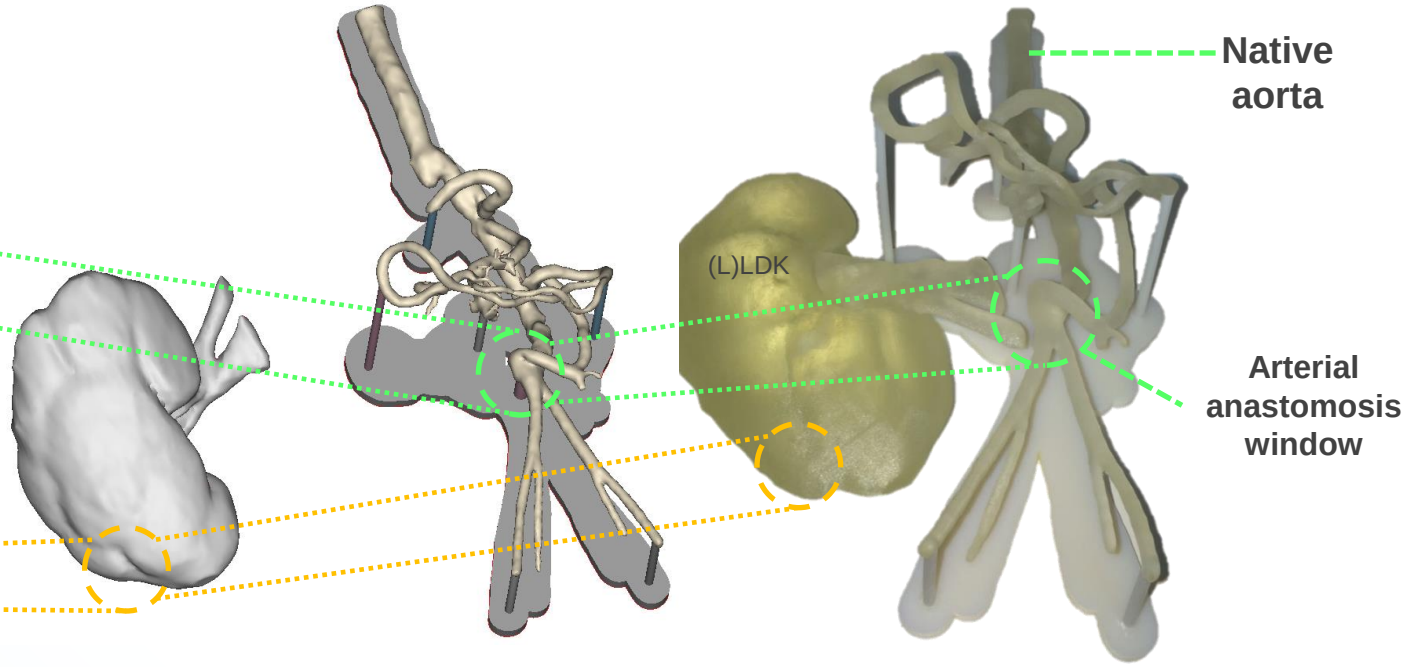




6 yr F, 18kg





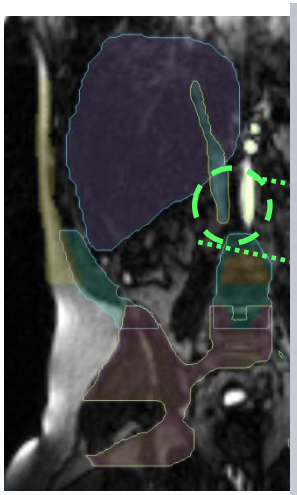


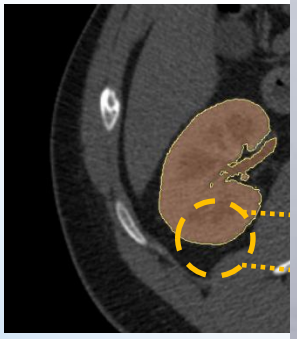
Geometrical correlation between CT/MR *vs* Segmented design *vs* model

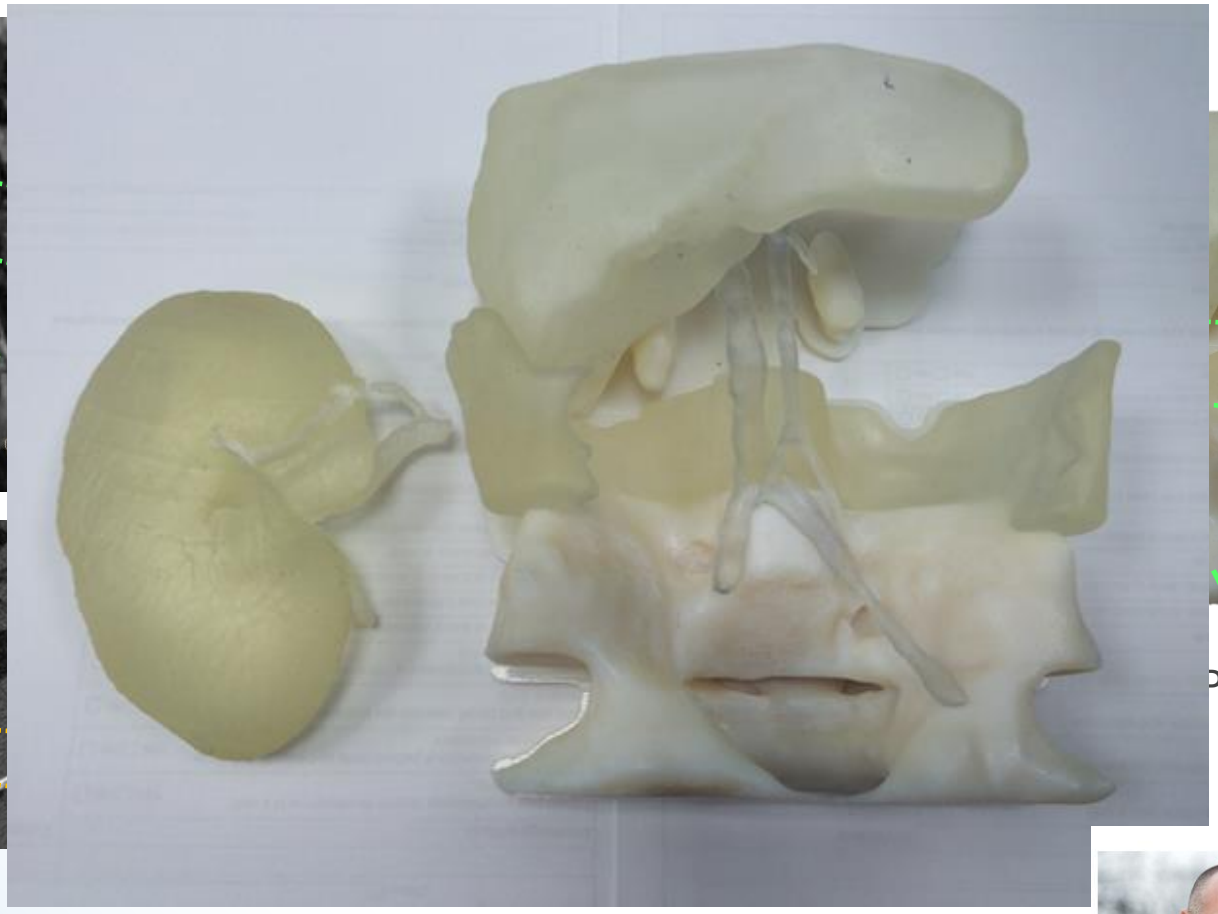
5 independent surgeons confirmed value as a preoperative planning tool (= 5)

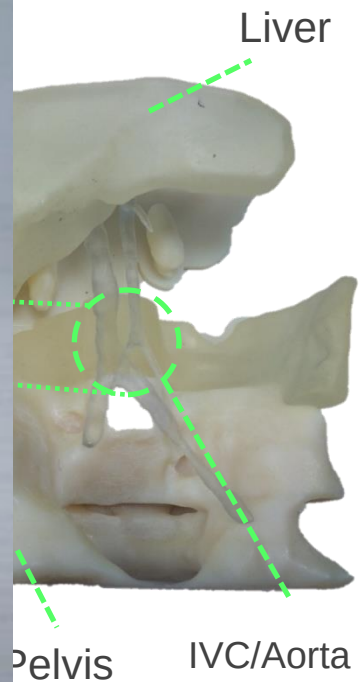
Case 1

Guy's and St Thomas' NHS
NHS Foundation Trust







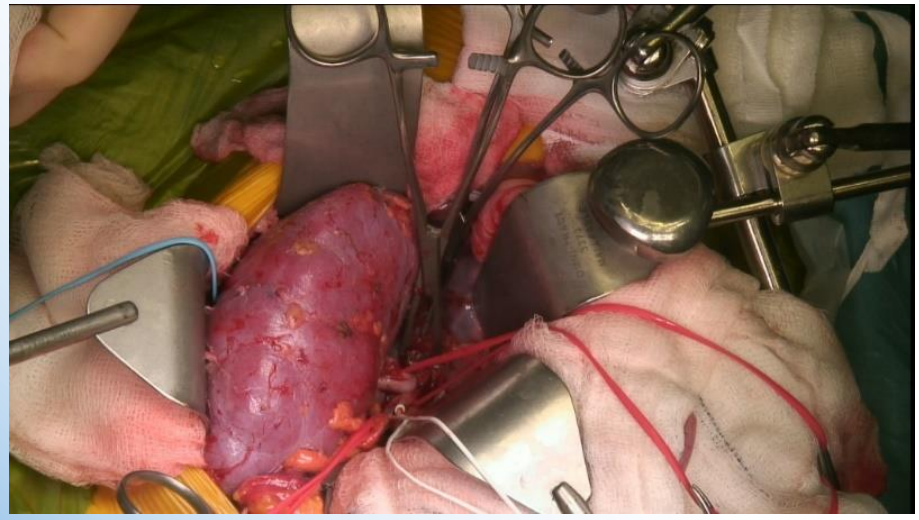


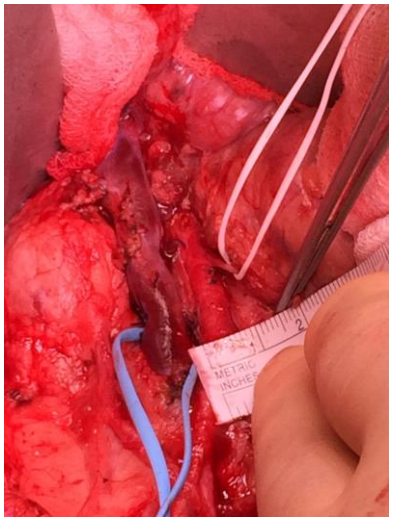
2 yr F, 10 kg,

previous laparotomies for bowel ischaemia

Surgeon:
=5 for planning and anatomy correlation;
=4 for kidney placement

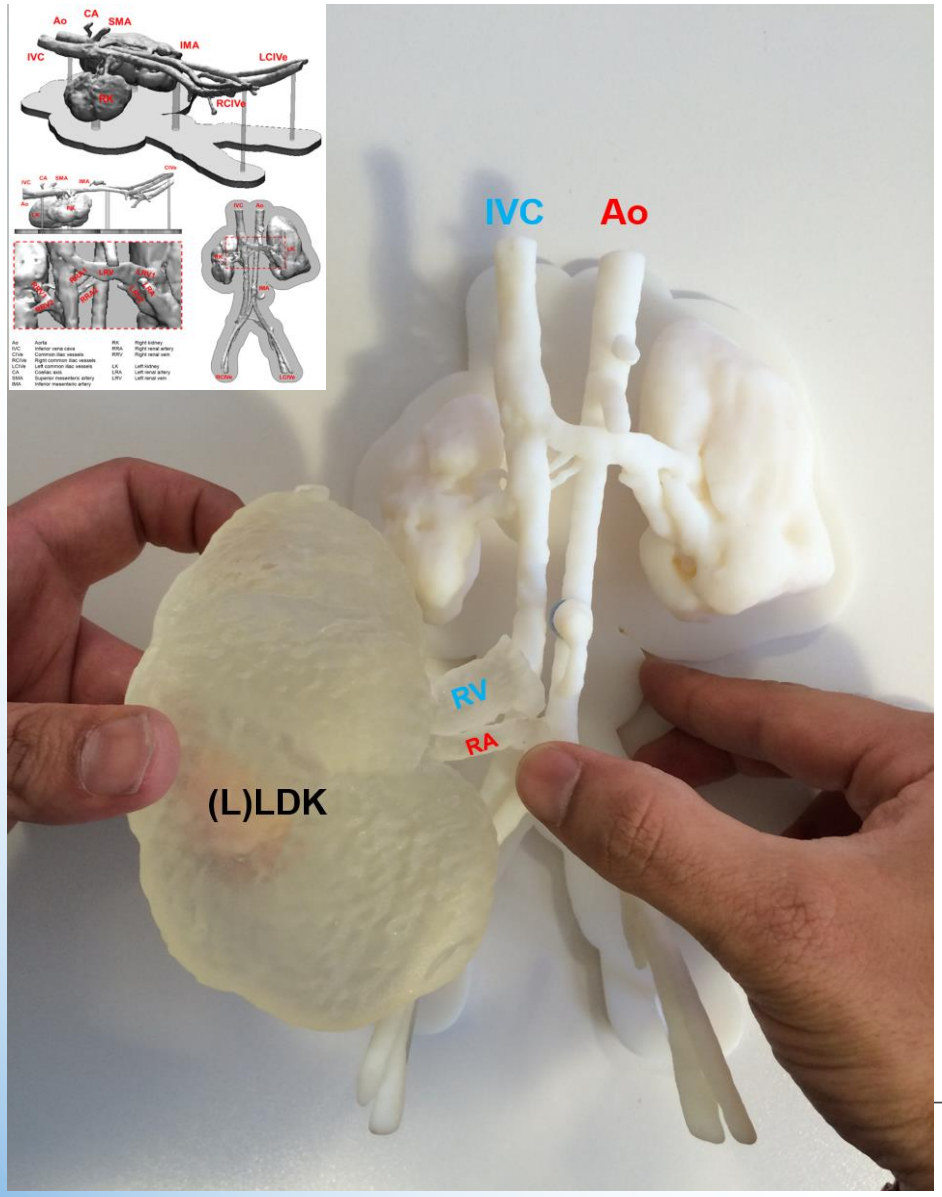
Family consenting
=5

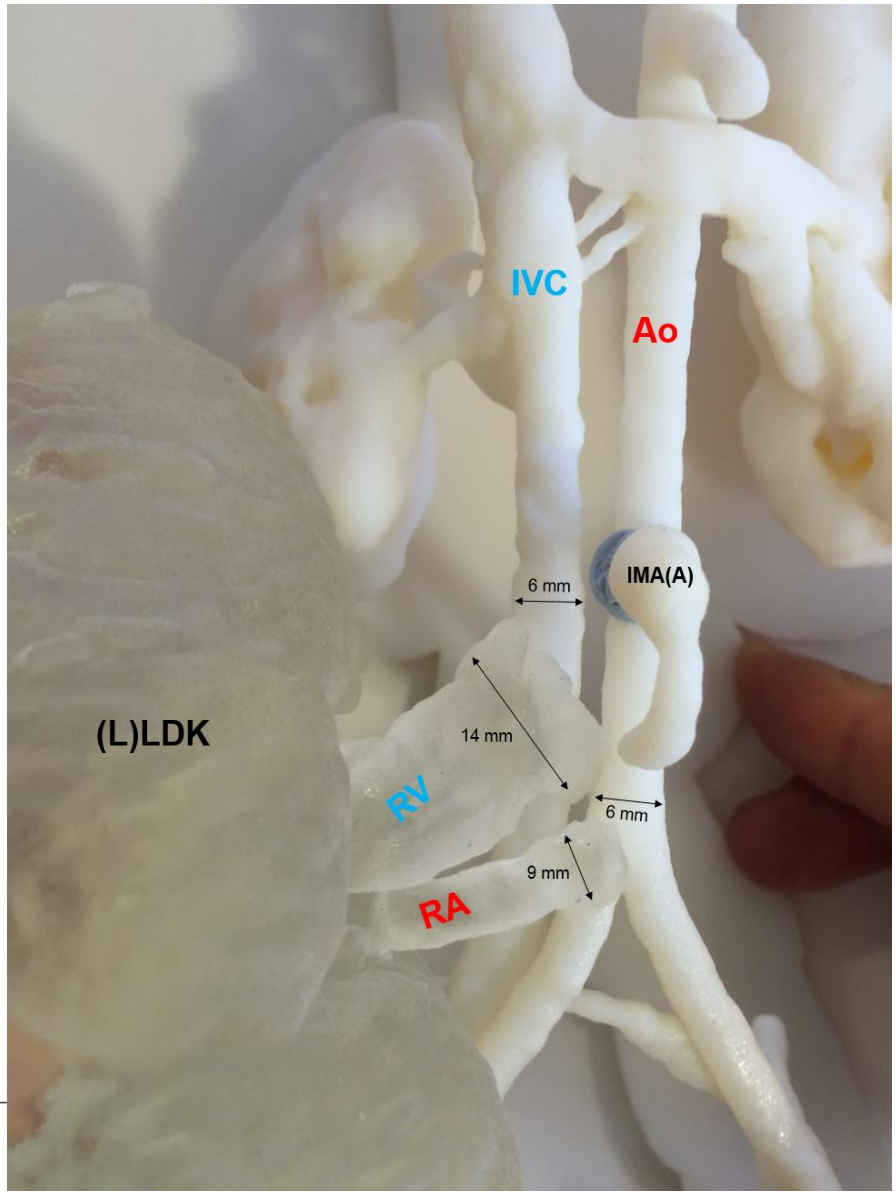




Case 2:
12 kg child , Renal and IMA aneurysms

Guy's and St Thomas' NHS
NHS Foundation Trust

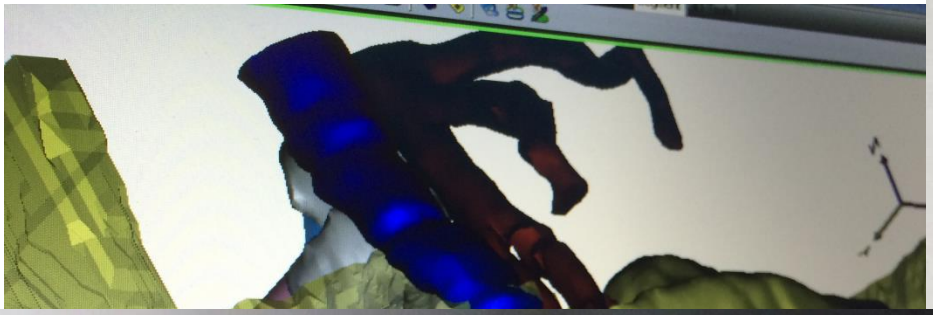


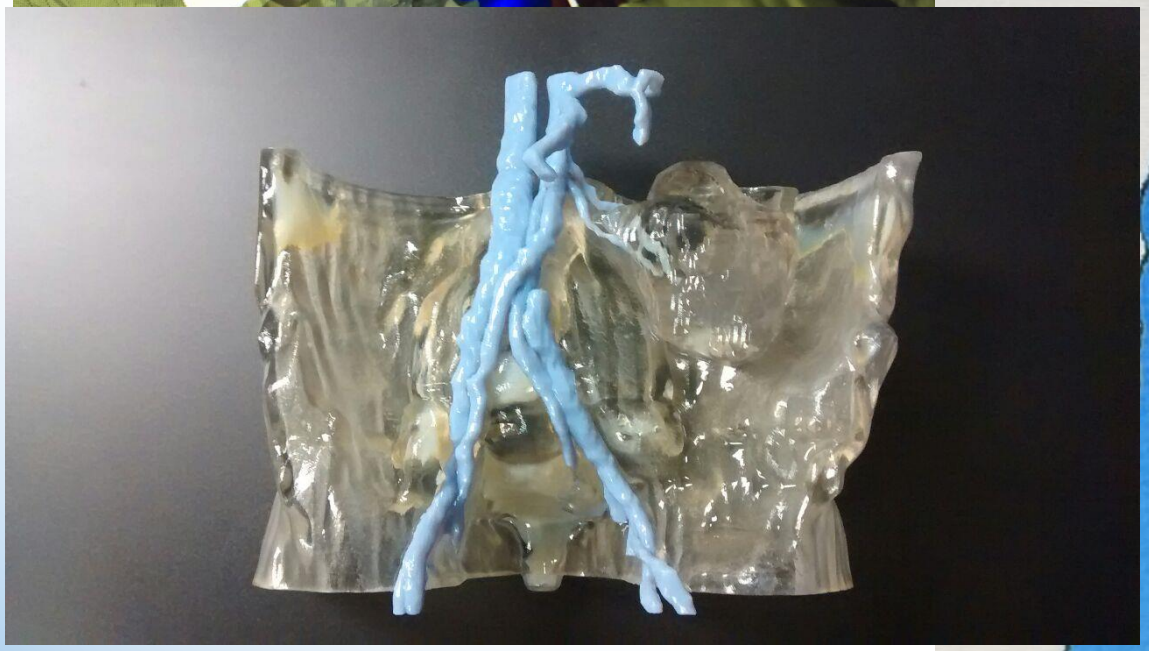


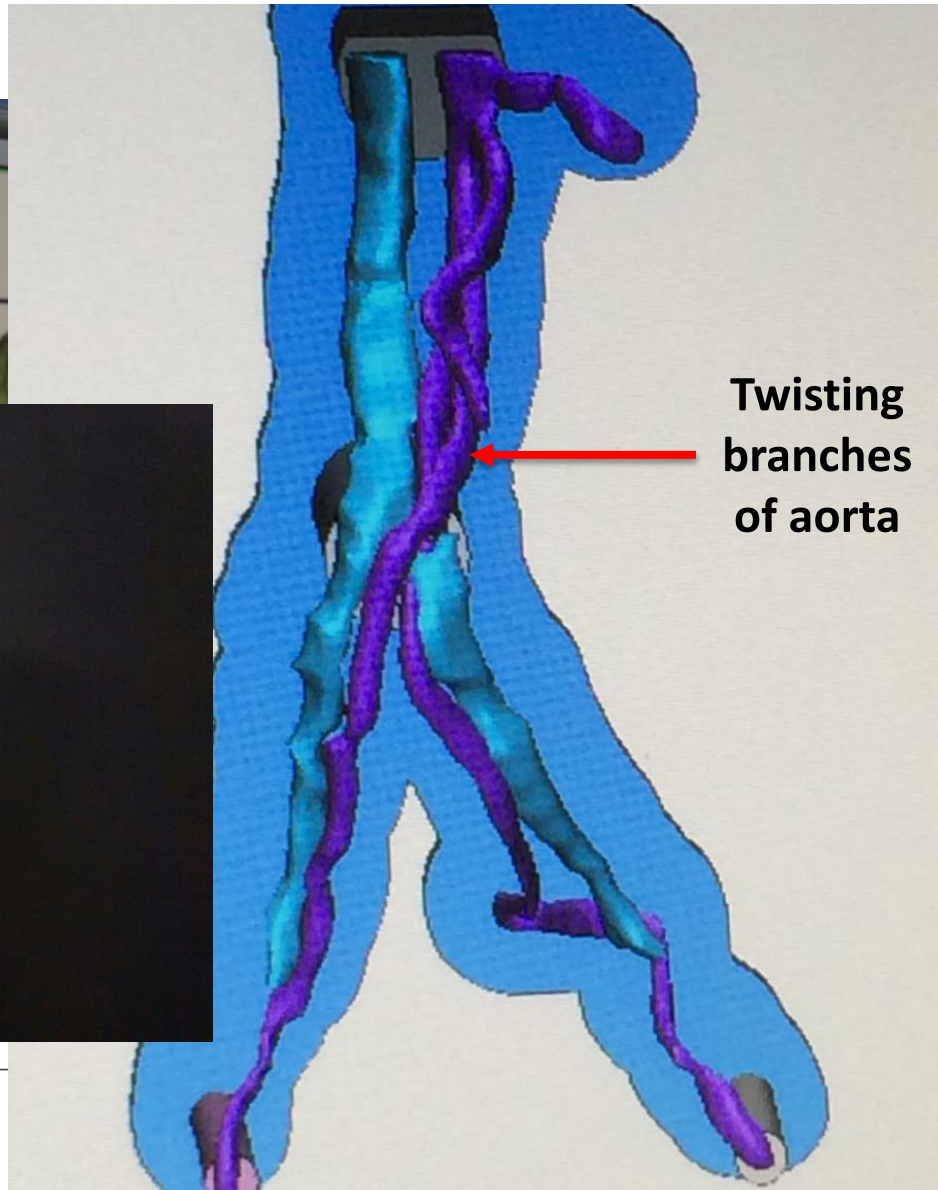
Case 3

14kg F, twisting branches of aorta with high divide

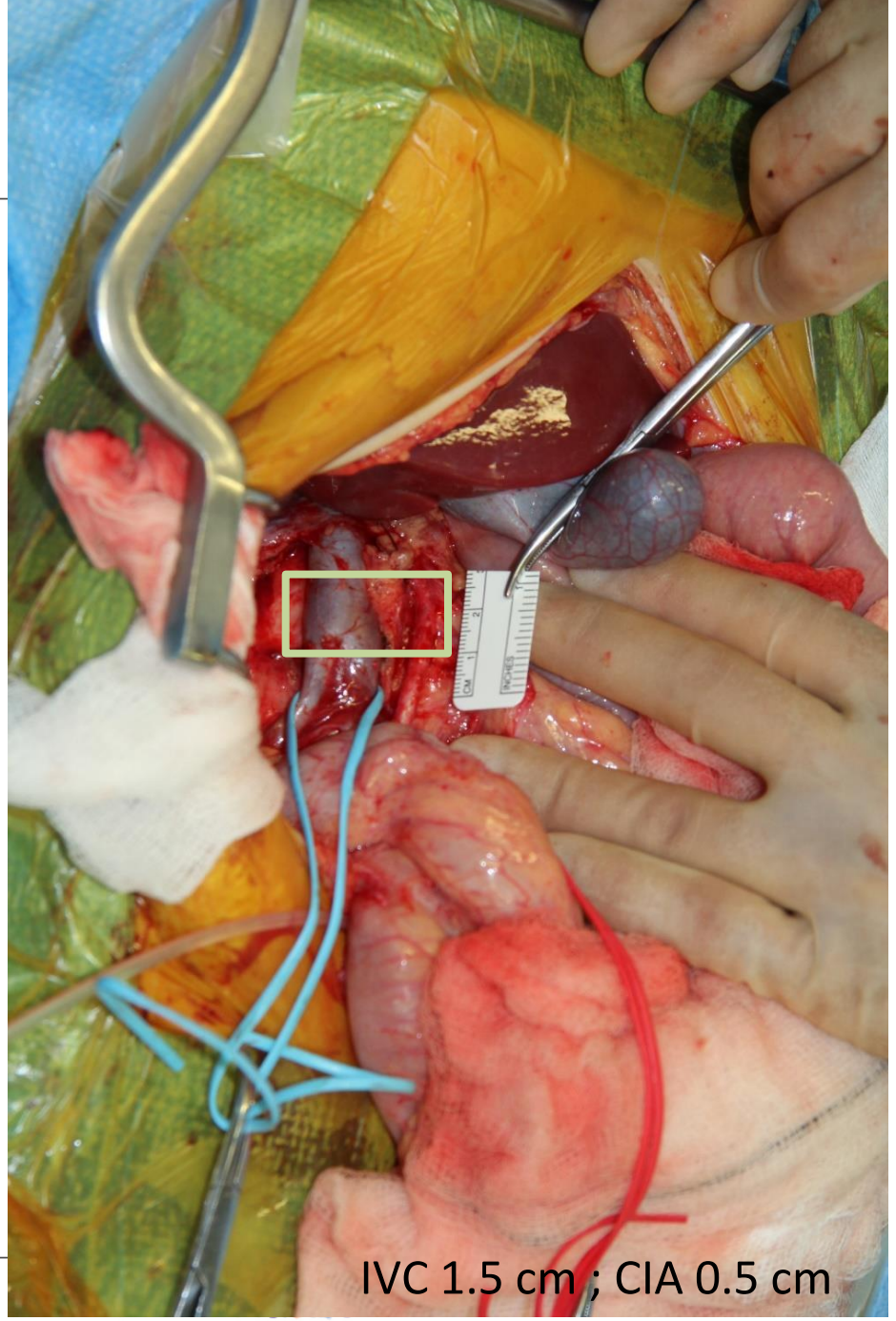
Guy's and St Thomas' NHS
NHS Foundation Trust








Ao
IVC


IVC 1.5 cm ; CIA 0.5 cm

Considerations for transplantation

Multidisciplinary discussion

- **Radiology, Nephrology, Tx Surgery, Anaesthetist, Vascular surgeon, Liver surgeon**

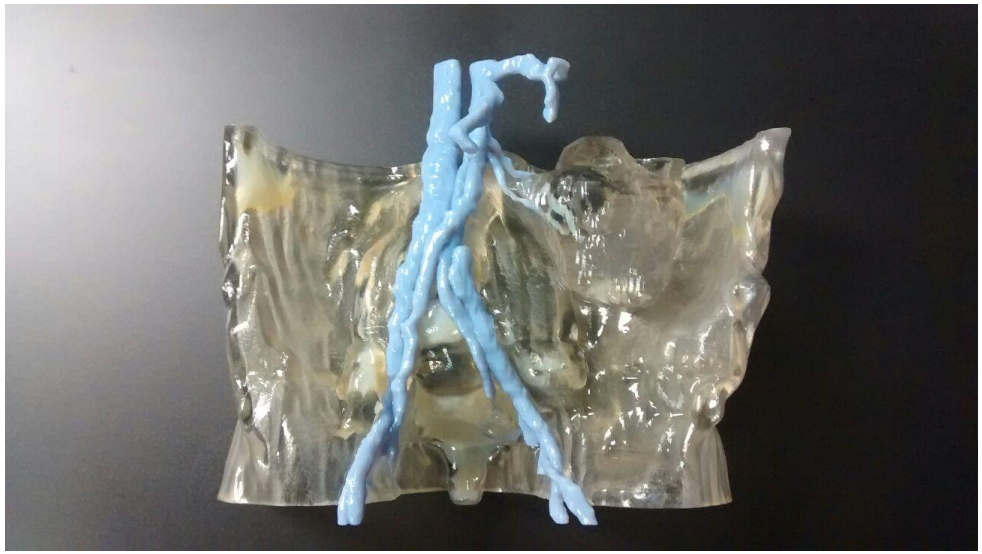
2 consultant surgeons

Prolonged anaesthesia- PICU

Inspection of vessels

Use of vascular conduits

Use of 3D printing



4th PAEDIATRIC KIDNEY TRANSPLANTATION SYMPOSIUM

A Multi-professional Meeting

6th December 2018 13:00-17:00 and
7th December 2018 9:00-17:00
London, UK

www.guystransplant.wordpress.com

Key Topics

Transplantation in small children

Pre-emptive transplantation – how to do it

Combined liver and kidney transplantation

Pancreas and islet cell transplantation in children

Auto-transplantation as treatment for reno-vascular hypertension

Paediatric transplant recipient in 21st century

Intraoperative management – what really matters

ABO and HLA incompatible transplantation

Limits in paediatric transplantation

Complex case exchange

Best abstract prize presentation

Guest speaker TBC

RCPCH has approved this activity for CPD in accordance with the current RCPCH CPD Guidelines

Registration

Consultants £90 (£70 before 20th November)
Trainees £70 (£50 before 20th November)
Nurses/Coordinators £20
jelena.stojanovic@doctors.org.uk

Organizing committee

Dr Jelena Stojanovic
Mr Nicos Kessaris
Professor Nizam Mamode

Guy's, Evelina, Great Ormond Street Hospitals teams:

P Chandak,N Kessaris, SD Marks, J Stojanovic, G Walsh, N Ware

3D Printing:

N Byrne, A Coleman,
N Karunanithy, J Carmichael,

GUY'S & ST THOMAS' CHARITY

Kind permission from all the patients and families

KING'S HEALTH PARTNERS