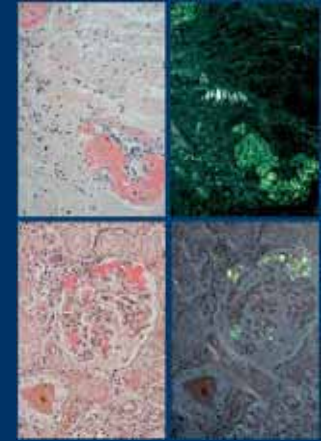




Eröffnungssymposium
des Amyloidose-Zentrums
Heidelberg

Samstag, 2. Mai 2009



UniversitätsKlinikum Heidelberg

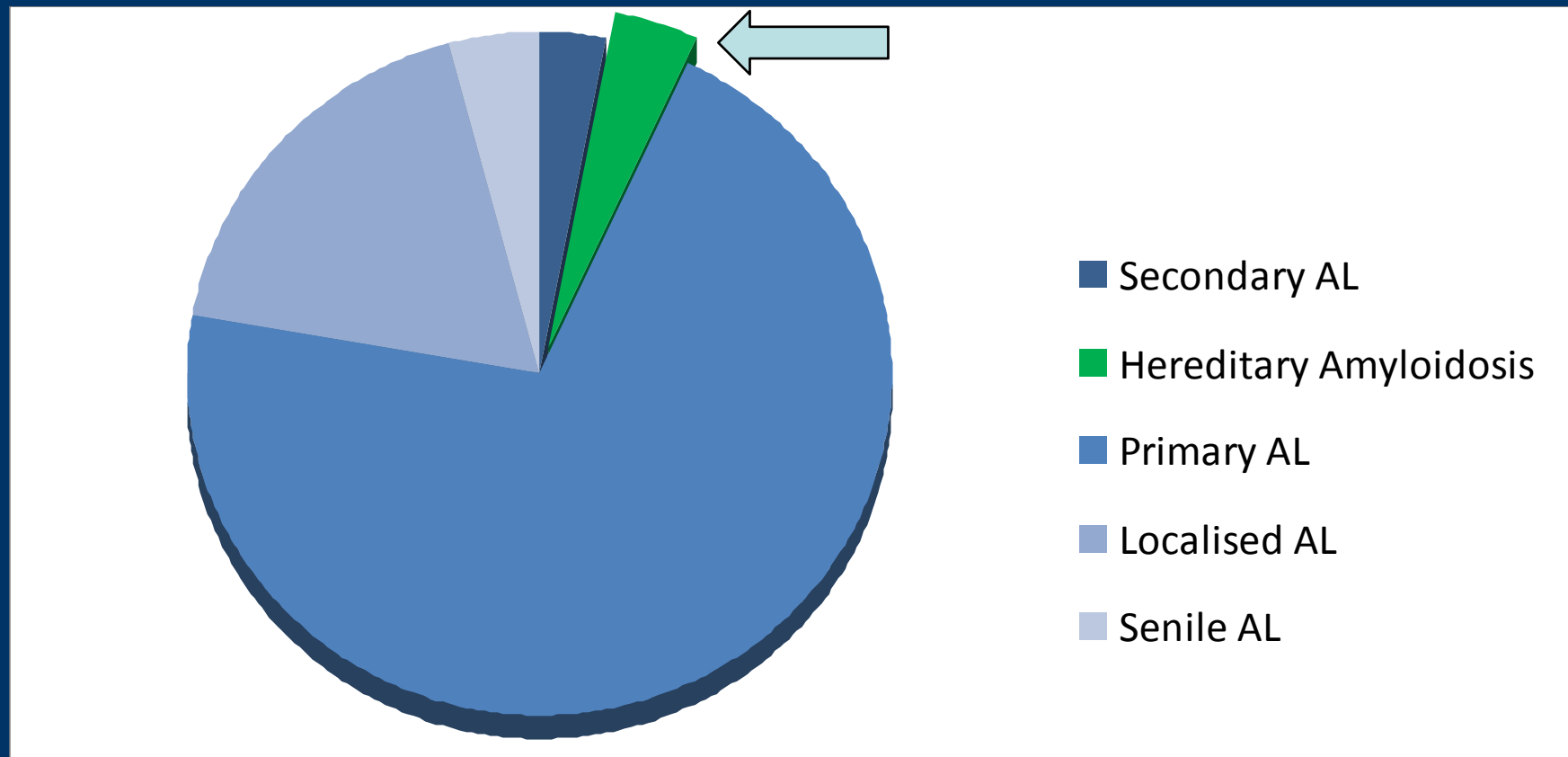
Amyloidosis

The (Transplant-) Surgeon's View

P. Schemmer



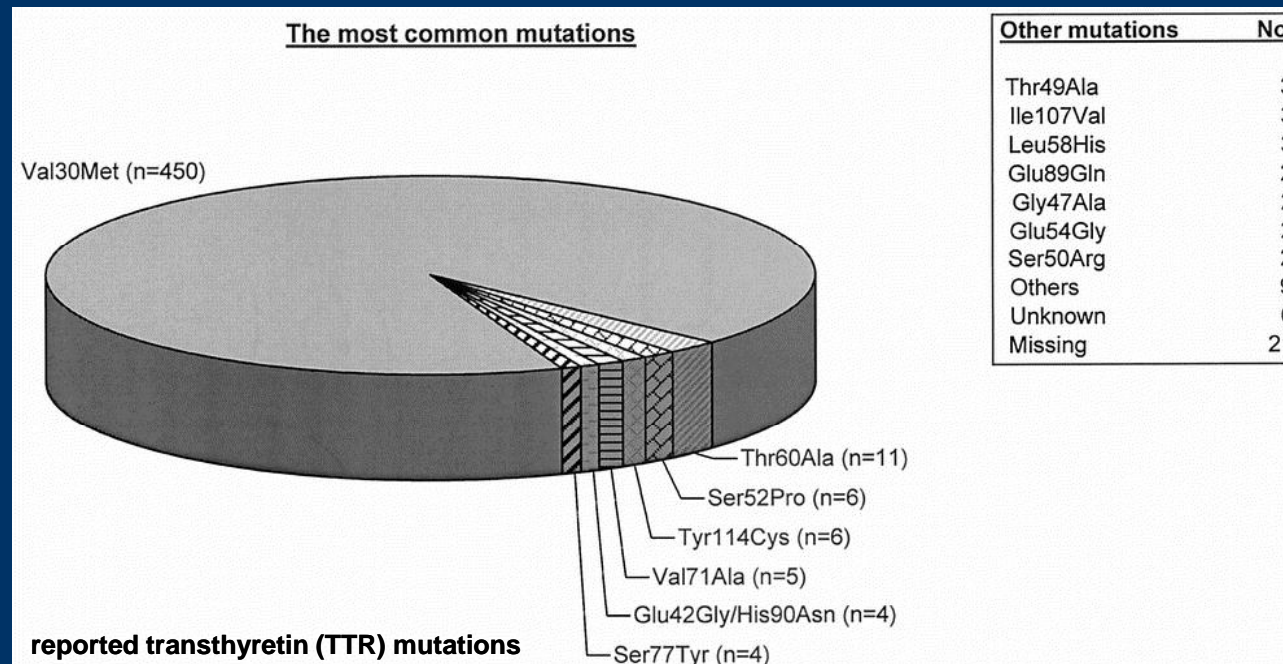
Classification of Amyloidosis





ATTR_{Val30Met}

- Familial amyloidotic polyneuropathy (FAP)
- Substitution of valine for methionine (Val30Met) is the most common
- Deposition of amyloid fibers in the extracellular matrix of Peripheral nerves, GI tract, Heart, Kidneys, Autonomic nervous system





Concept of LTx in FAP

Mutant TTR (transthyretin amyloidosis) is produced mainly
in the liver



Orthotopic LTx eliminates the source of the defect
TTR molecule
(not ApoA I + ApoA II)



FAPWTR (2008-06-30)

- **N = 1455**

Portugal (N = 650)

France (N = 192)

Sweden (N = 119)

Germany (N = 50)

Mainz 20

Hannover 17

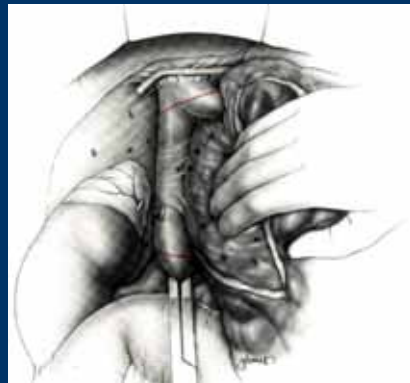
Heidelberg 11

Freiburg 1

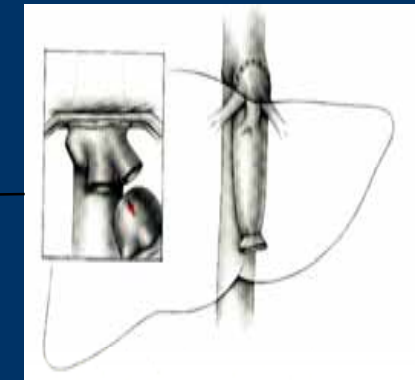
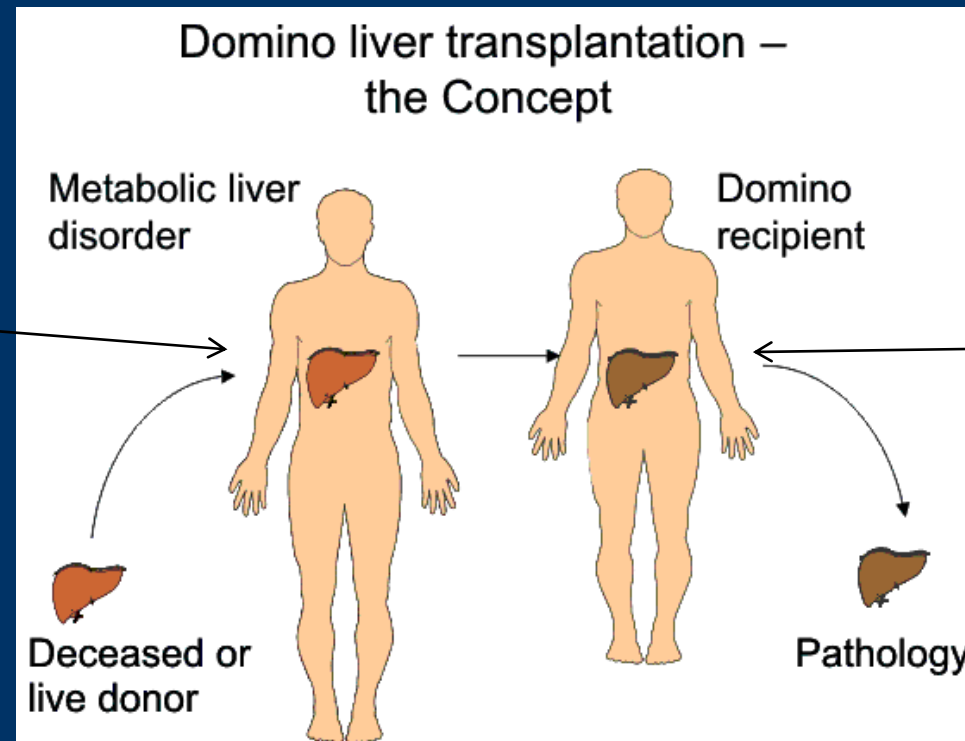
Münster 1



Domino-LTx



conventional

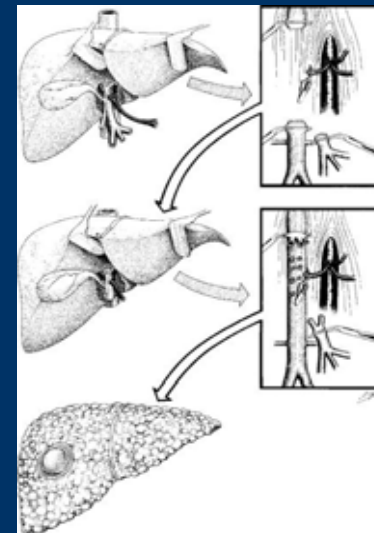


piggy-back



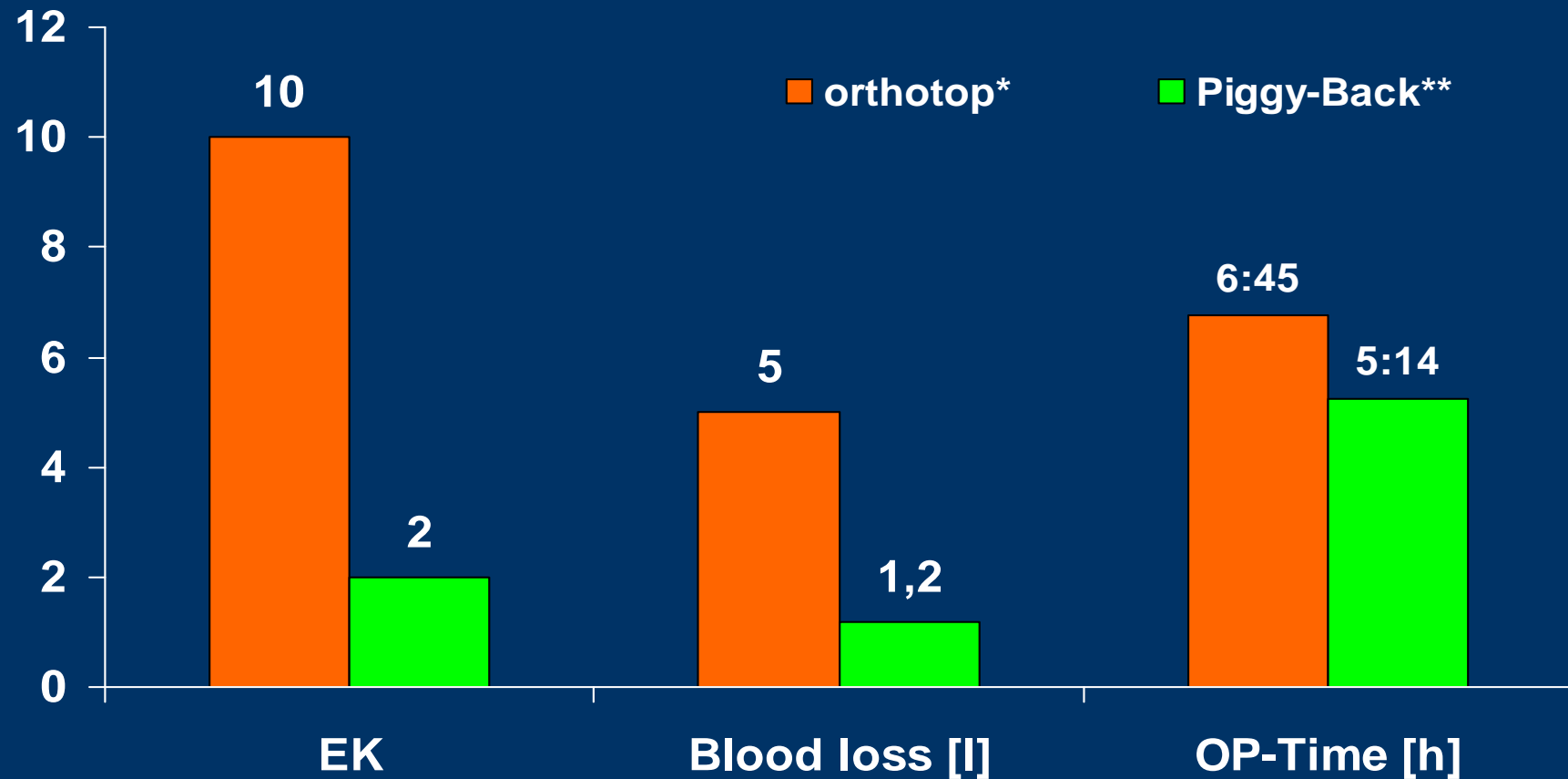
Operative Procedure

- long resection of retrohepatic caval vein
- hemodynamic tolerance test → venous bypass if necessary
- achieve long arterial and portal segments for graft and patient
- Preservation of the inferior vena cava





conventional vs Piggy-Back



*01.01.2001 bis 30.11.2001 (n = 27)

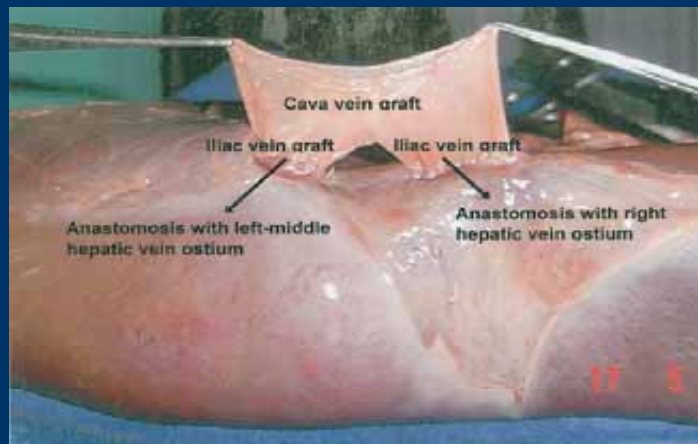
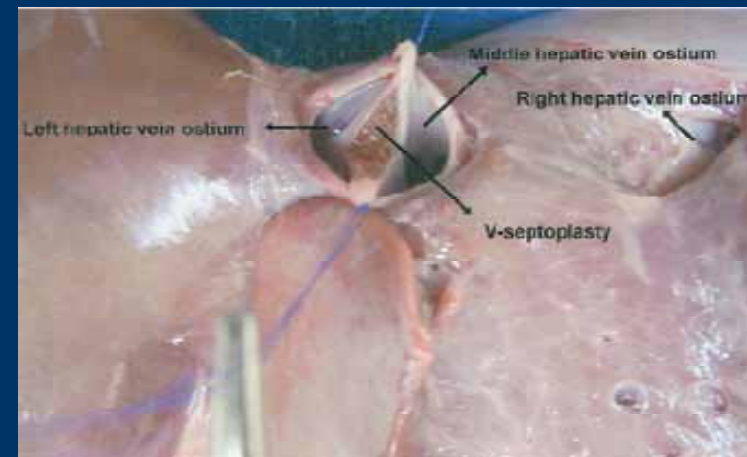
**01.12.2001 bis 31.3.2005 (n = 221)



Double Piggyback

Lacerda, Miranda, Amorim et al, Transplantation Proceedings 2008

- experimental procedure
- main technical problem is the length of the hepatic veins
- reconstruction of the vascular outflow using an iliac/cava graft



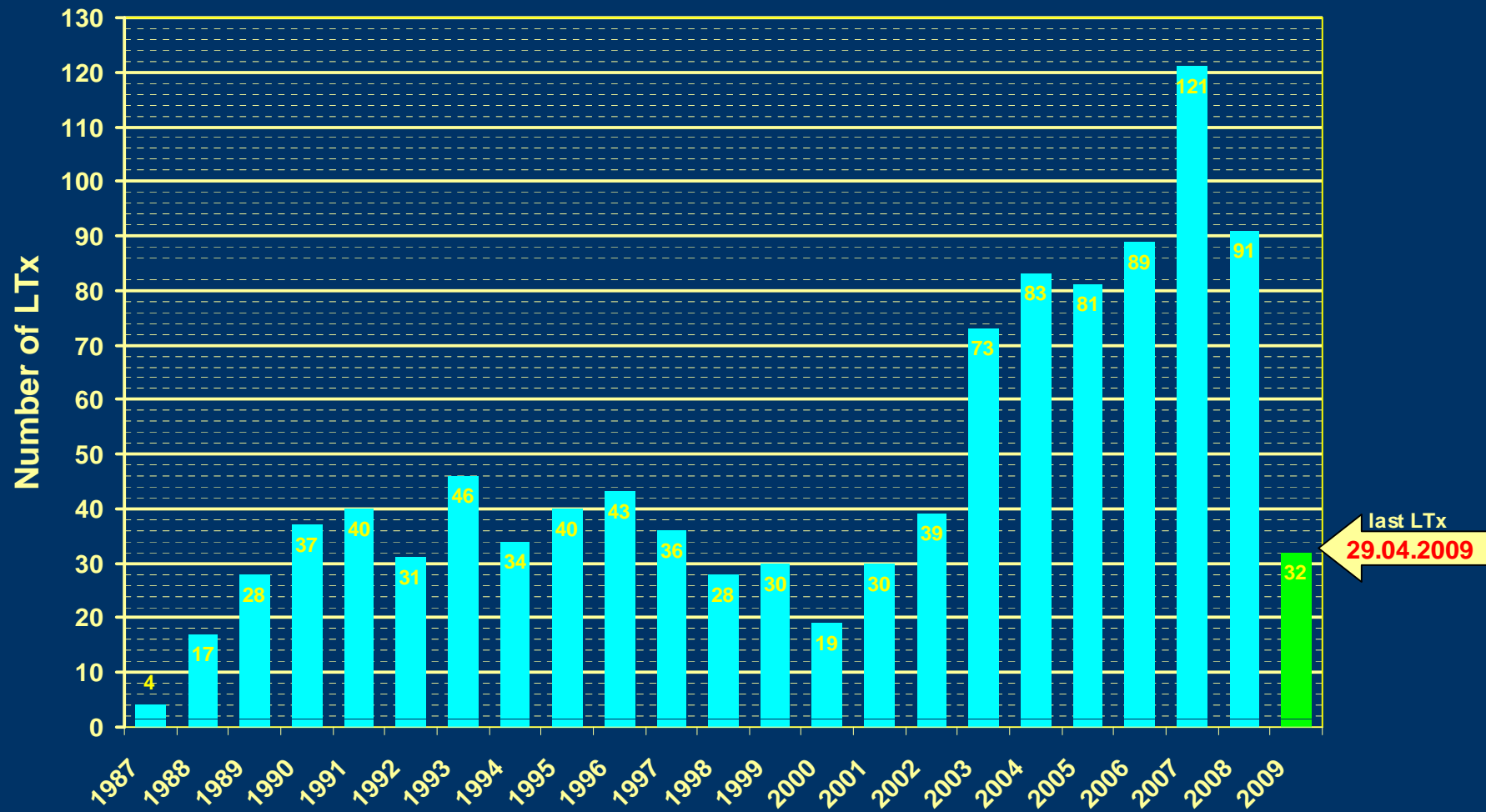
Reconstruction of venous outflow with a cava graft

FAP liver septoplasty between left / right hepatic vein



Liver Transplantation

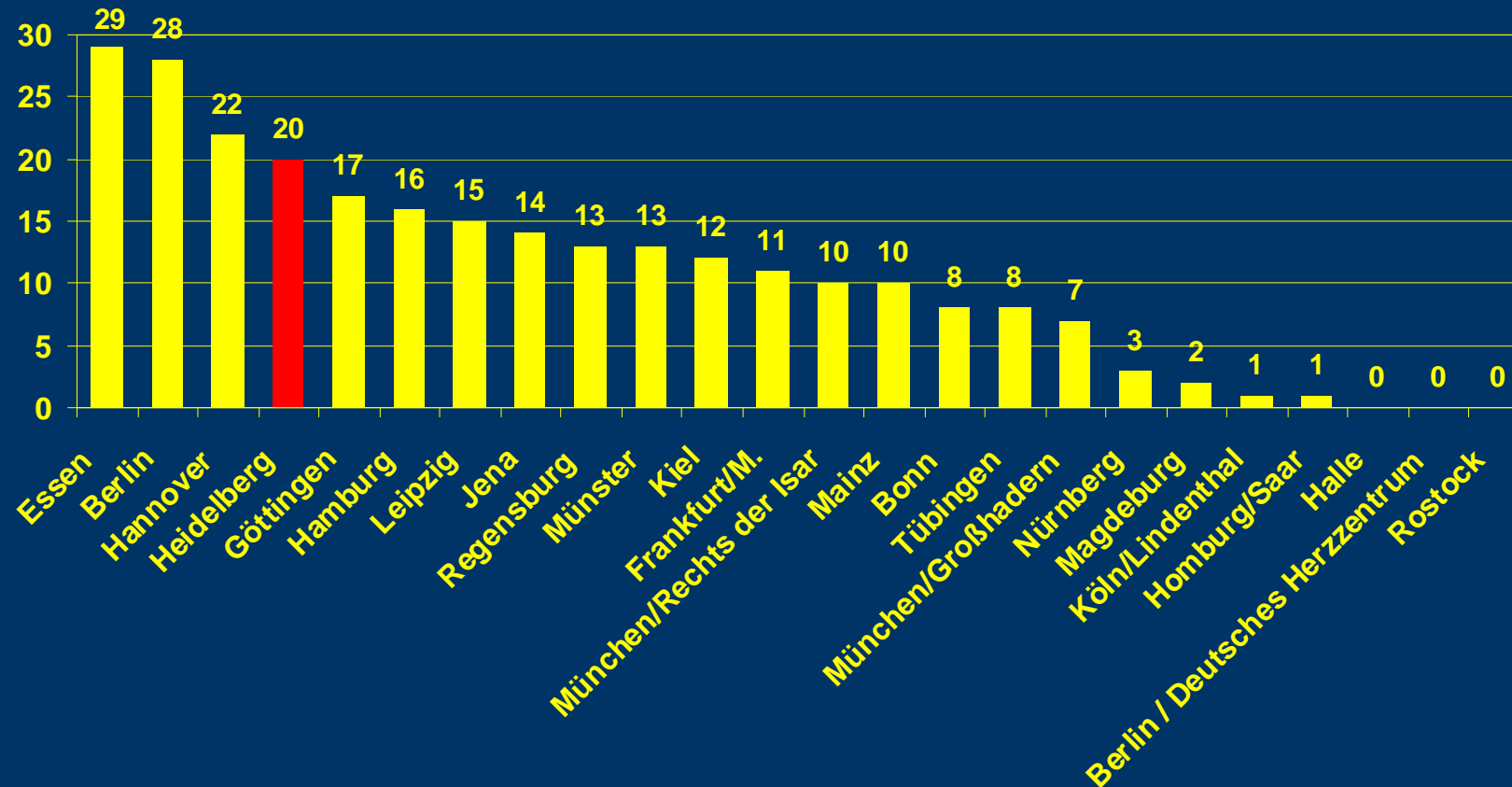
1987 – 29.04.2009 (n=1072)





Liver Transplantation* in Germany

(01.01. - 31.03.2009)

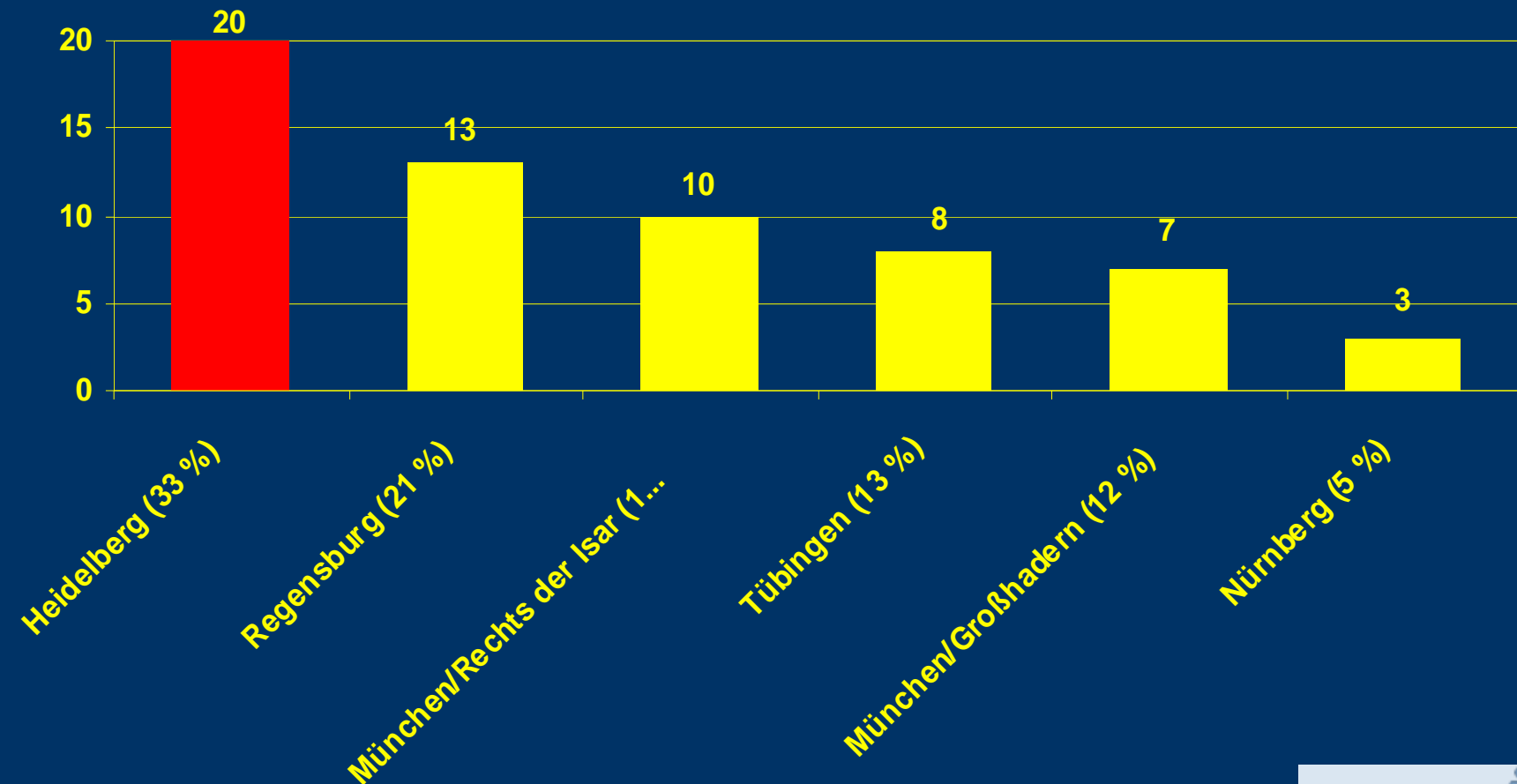


* postmortal donation without Split-Liver



Liver Transplantation in South-Germany

(01.01. - 31.03.2009)

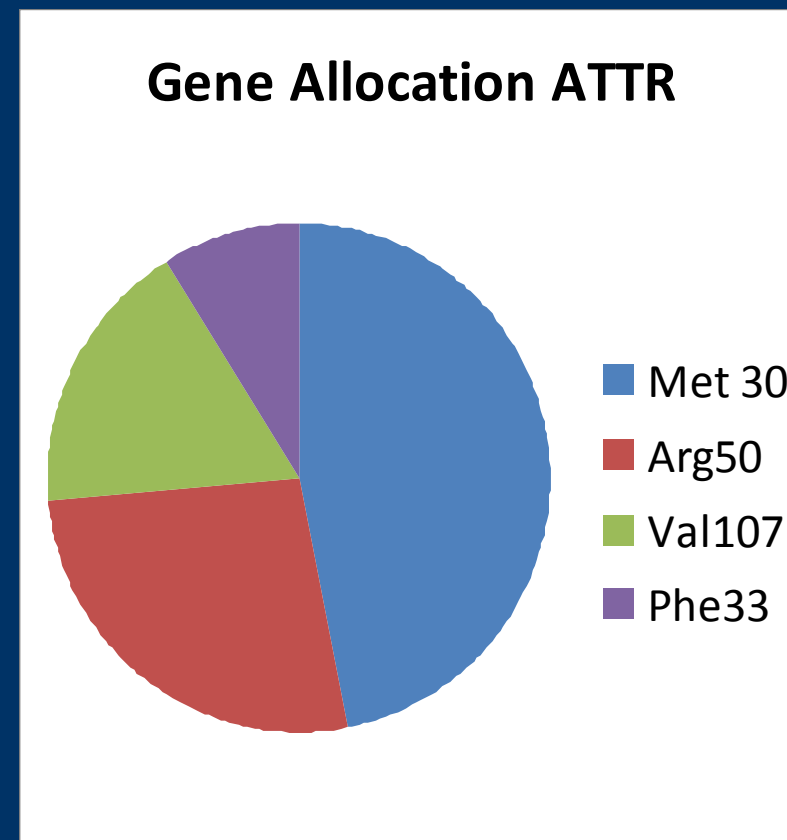


* postmortal donation without Split-Live



LTx ATTR_{Val30Met} in Heidelberg

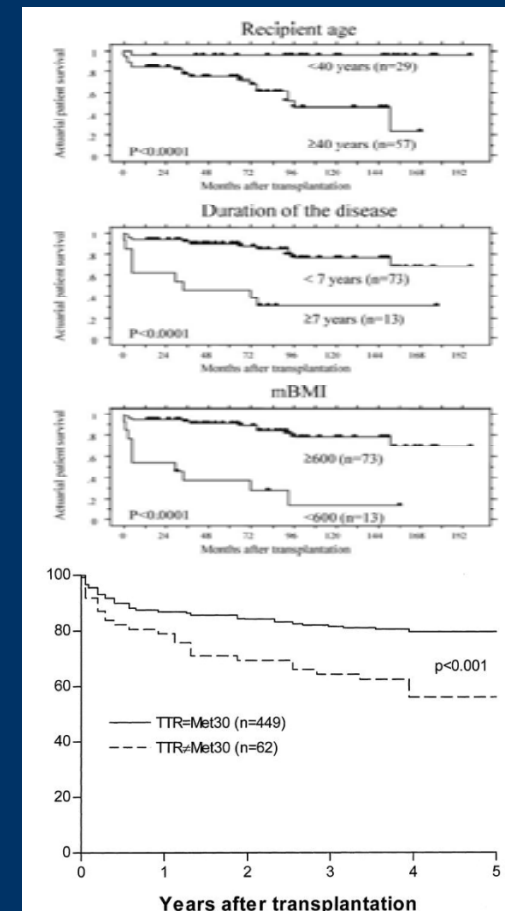
- N = 11
- Gender:
 - male: 7 (64%)
 - female: 4 (36%)
- Age: 49.5 (27 – 70)





LTx ATTR_{Val30Met} in Heidelberg

- **Domino LTx:** **N = 6 (55%)**
- **Follow-up period:** **3.9 y (0 – 8)**
- **survival rate:** **72%**
 - Met 30: **80%**
 - Non-Met30: **66%**



Herlenius A, et al. Transplantation
2004



Conclusion

- LTx is the only curative treatment for patients with familial amyloidotic polyneuropathy (FAP)
- If prognostic factors are not optimal, LTx should be considered well
- DLTx could extend the donor-pool