

# Surgical Considerations in the Management of Wilms Tumor

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**Disclosures: None.**

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# Wilms Tumor

- Most common renal tumor of childhood
- Only about 500-650 cases per year in United States
- Model for the rare disease that has had successful advancements in treatment and outcome owing to multicenter collaboration and thoughtful research
  - Surgical technique
  - Exquisite chemosensitivity
  - Patient risk stratification

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# Surgery is a Critical Component of Therapy

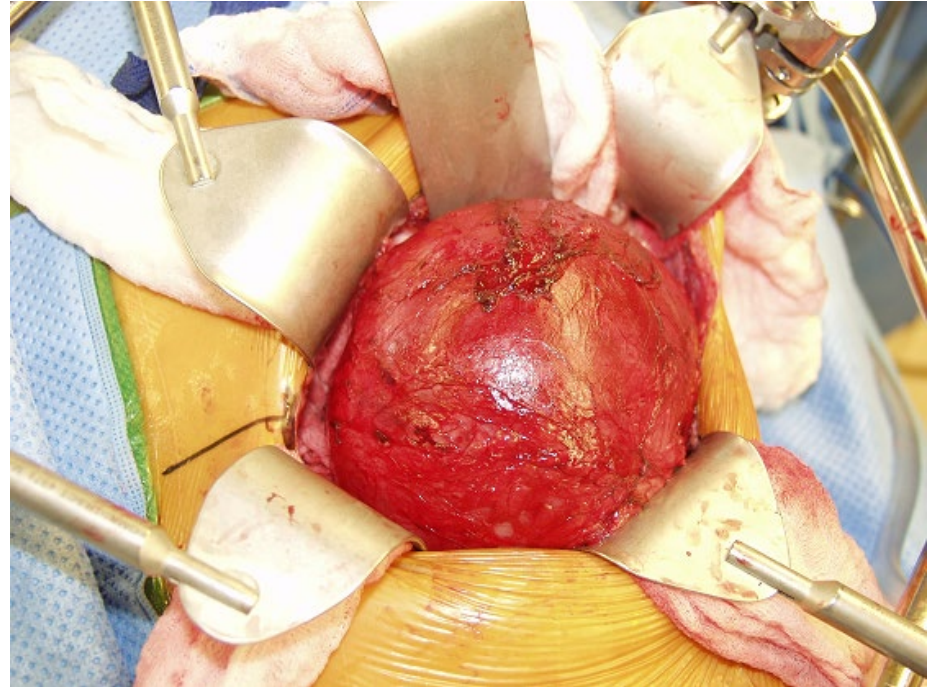
- Excision of tumor in kidney and in lymph nodes
- Avoiding spill and spread
- Surgical staging
- Preoperative planning will optimize surgical outcome

# Goals of Surgery for Wilms Tumor

- Goal is excision of the tumor in its entirety with proper staging and without spill
- Assessment of contralateral renal unit, collecting system, venous structures/right atrium
- Avoid spill (and distinguish it from rupture)
- Proper lymph node “sampling”

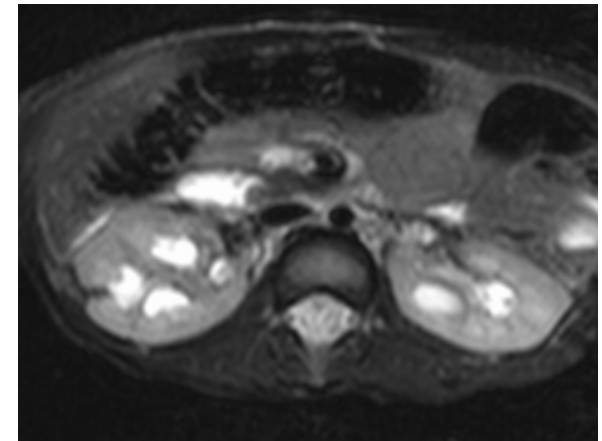
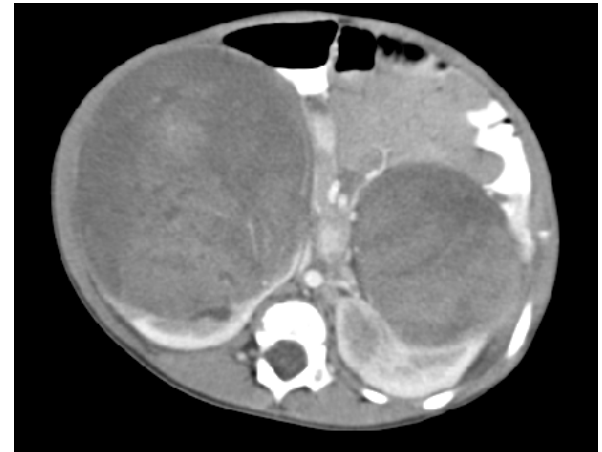
# Preoperative Planning

- Assess upfront resectability
- Assess venous and atrial tumor involvement
- Assess collecting system involvement
- Plan approach



# Assessment of Resectability

- Even large tumors can be resected in their entirety
- Left to surgeon's discretion
- Look for metastases and involvement of contralateral kidney
- In general, avoid removing adjacent organs other than the adrenal gland



# Looking for Metastases

- Isolated lung metastases: can still proceed with upfront nephrectomy
- Evaluate contralateral kidney with thin-slice cross-sectional imaging





**Stage IV FH  
WT (lung  
mets only)**

AREN  
0533

No LOH  
1p/16q

LOH 1p/16q

Lung CR by week  
6 of DD4A (VAD)

Incomplete lung  
response by week  
6 of DD4A

Continue DD4A  
NO lung rads

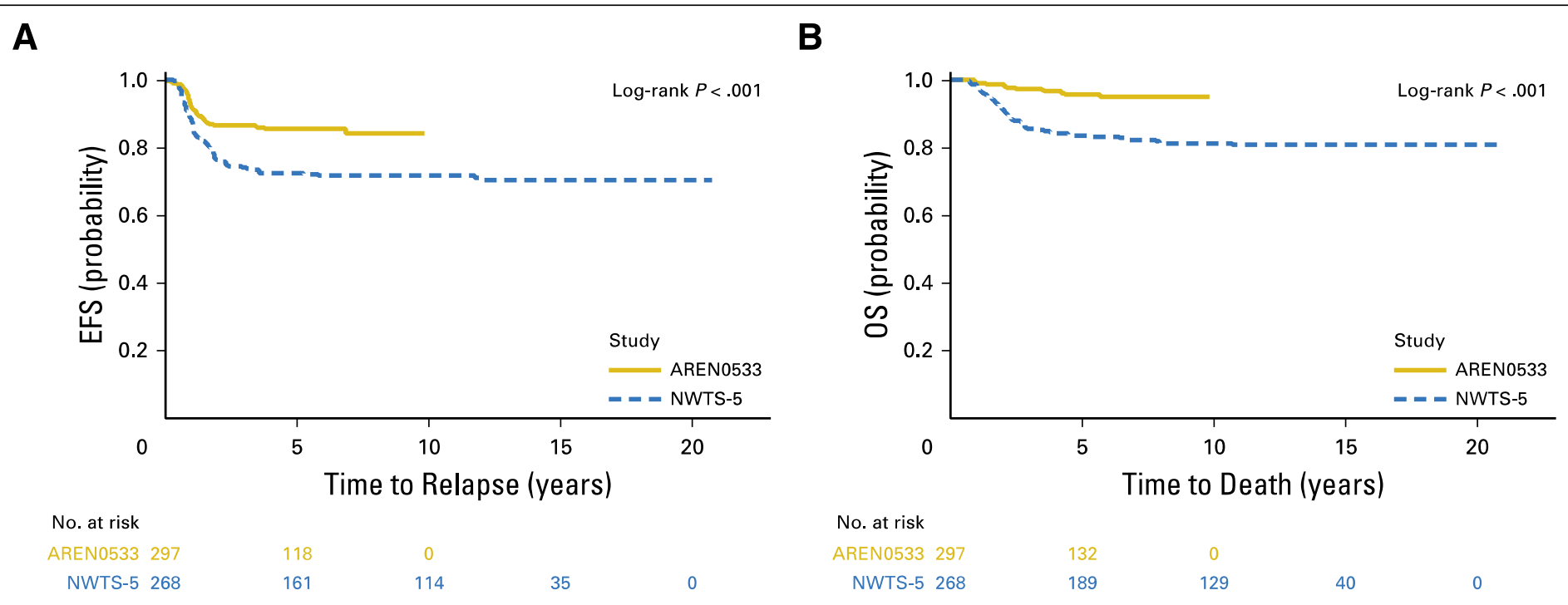
Regimen M  
(5 drugs) + lung  
rads



# Treatment of Stage IV Favorable Histology Wilms Tumor With Lung Metastases: A Report From the Children's Oncology Group AREN0533 Study

David B. Dix, Nita L. Seibel, Yueh-Yun Chi, Geetika Khanna, Eric Gratas, James R. Anderson, Elizabeth A. Mullen, James I. Geller, John A. Kalapurakal, Arnold C. Paulino, Elizabeth J. Perlman, Peter F. Ehrlich, Marcio Malogolowkin, Julie M. Gastier-Foster, Elizabeth Wagner, Paul E. Grundy, Conrad V. Fernandez, and Jeffrey S. Dome

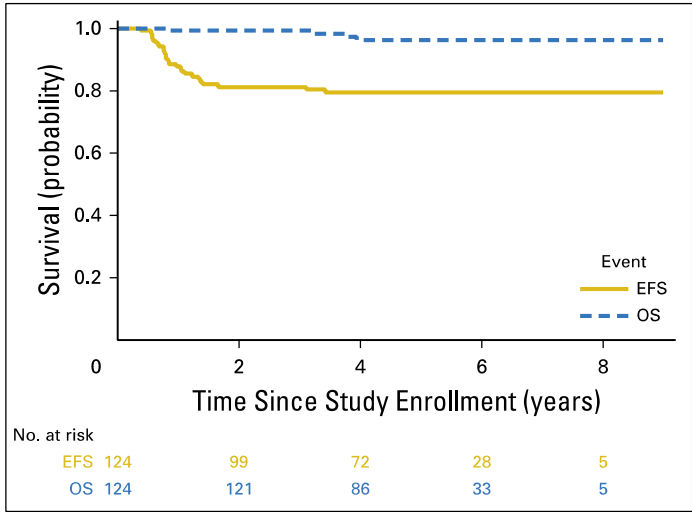
JCO 2018



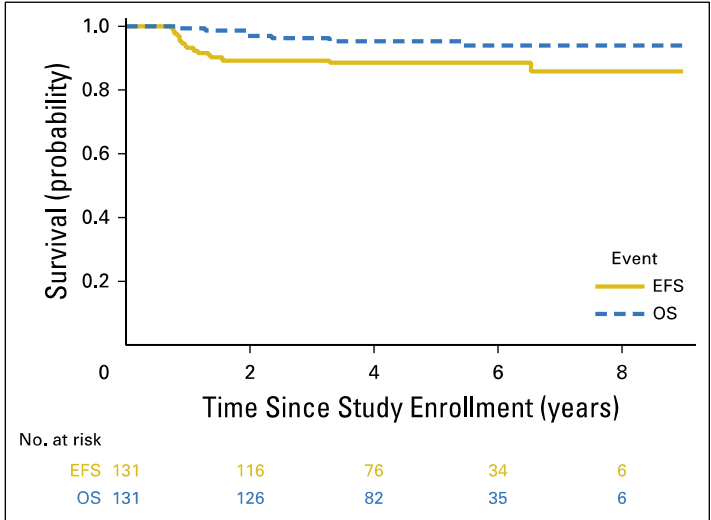
**Fig 5.** (A) Event-free survival (EFS) and (B) overall survival (OS) for patients with isolated pulmonary metastases in AREN0533 compared with NWTS-5.

CR

IR



**Fig 3.** Event-free survival (EFS) and overall survival (OS) for patients with lung nodule complete response who completed treatment with vincristine/dactinomycin/doxorubicin without lung radiation therapy.



**Fig 4.** Event-free survival (EFS) and overall survival (OS) for patients with incomplete lung nodule response without loss of heterozygosity who completed treatment with lung radiation therapy and four cycles of cyclophosphamide/etoposide in addition to vincristine/dactinomycin/doxorubicin.

# Different Types of Transperitoneal Incisions

- Midline
- Chevron
- Makuuchi
- Thoracoabdominal
- Not extraperitoneal

# Assessment of Contralateral Kidney

- Preoperative imaging (thin-slice cross-sectional imaging) is generally sufficient to evaluate contralateral renal unit
  - Lesions missed on CT tended to be <2 cm
  - Less risk with newer CT protocols
- If concerns, can palpate contralateral kidney intraoperatively before planned nephrectomy

FATE OF BILATERAL RENAL LESIONS MISSED ON PREOPERATIVE  
IMAGING: A REPORT FROM THE NATIONAL WILMS TUMOR STUDY  
GROUP

MICHAEL L. RITCHEY,\* ROBERT C. SHAMBERGER, THOMAS HAMILTON, GERALD HAASE,  
PEDRAM ARGANI AND SUSAN PETERSON



Seattle Children's®  
HOSPITAL • RESEARCH • FOUNDATION

Ritchey 2005; Ritchey 2001

# Assessment of Thrombus

	Sensitivity	Specificity
Primary Nephrectomy		
CT	65.6%	84.8%
Doppler US	45.8%	95.7%
Secondary Nephrectomy		
CT	86.7%	90.6%
Doppler US	66.7%	100%

← all thrombi

cavoatrial thrombi →

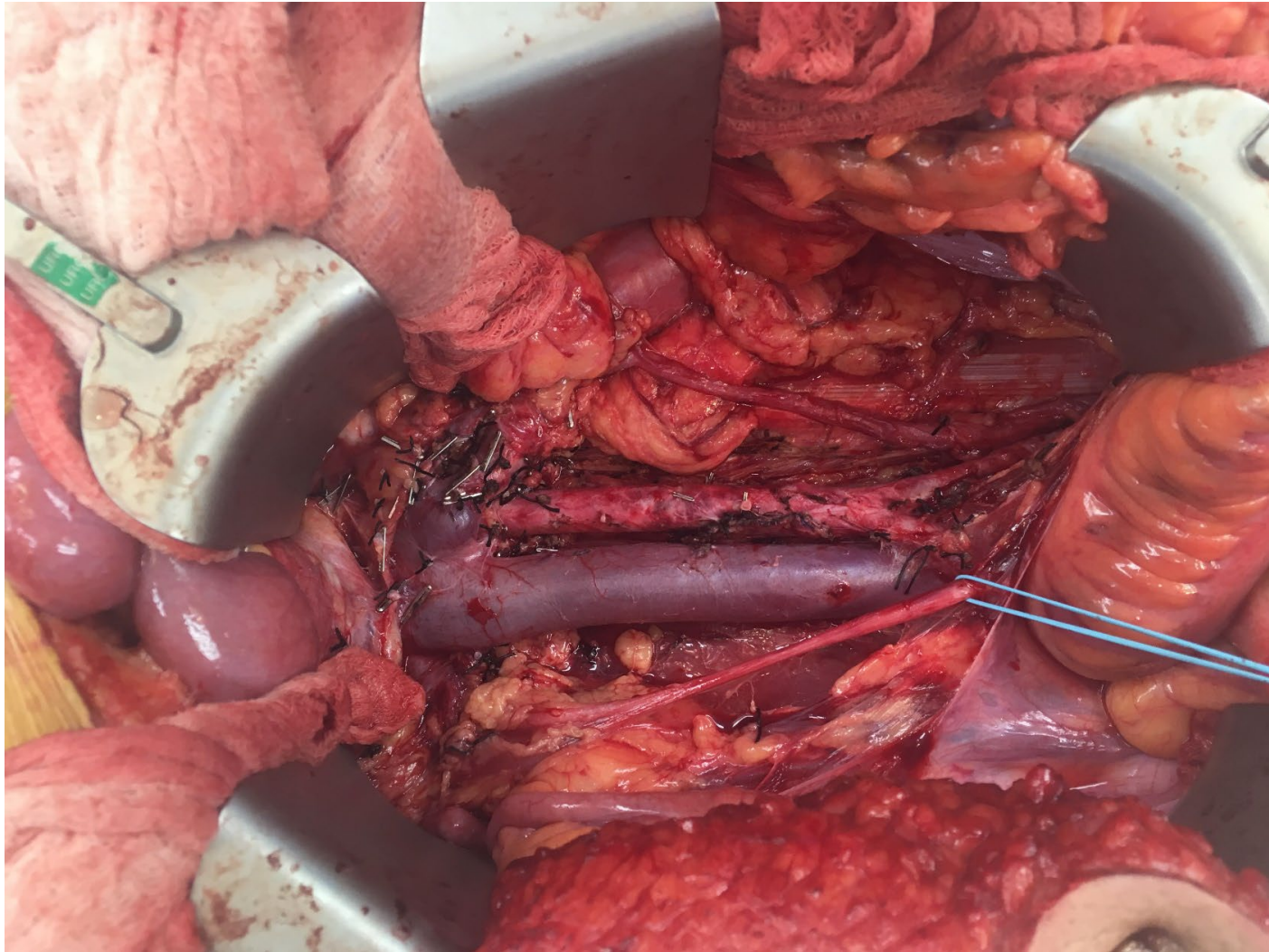
	Sensitivity
Primary Nephrectomy	
CT	84.6%
Doppler US	70.0%
Secondary Nephrectomy	
CT	96.0%
Doppler US	68.8%

CT correctly predicted extent of tumor in 80.9% of primary nephrectomy cases and 88.5% of secondary nephrectomy cases

# Vertical Midline

- Advantages
  - Modifiable to reach higher and lower
  - Approach to LNs both cranially and caudally
  - Can potentially use for bilateral tumor
  - Can combine with sternotomy for high level venous tumor thrombi
- Disadvantages
  - Poor exposure to upper-lateral retroperitoneum

# Vertical Midline

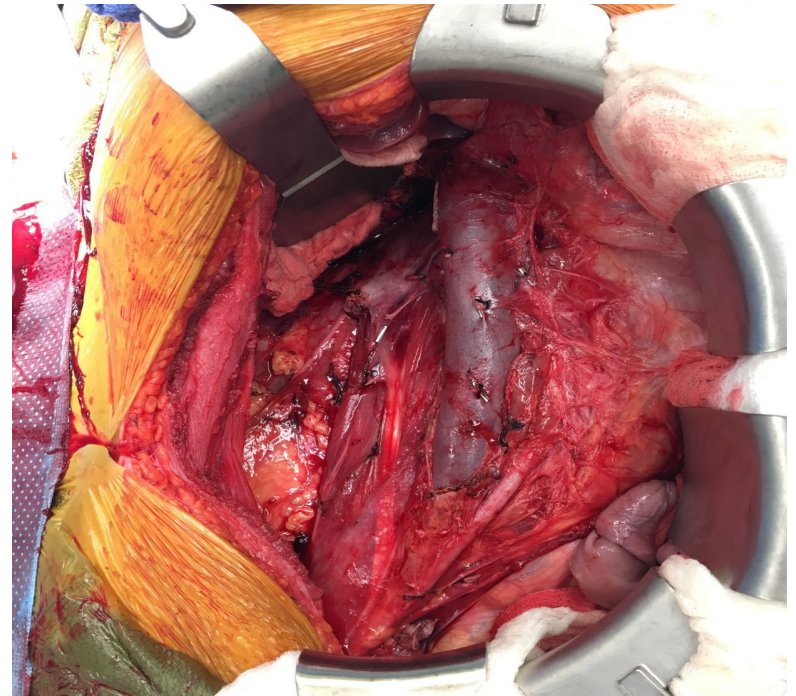
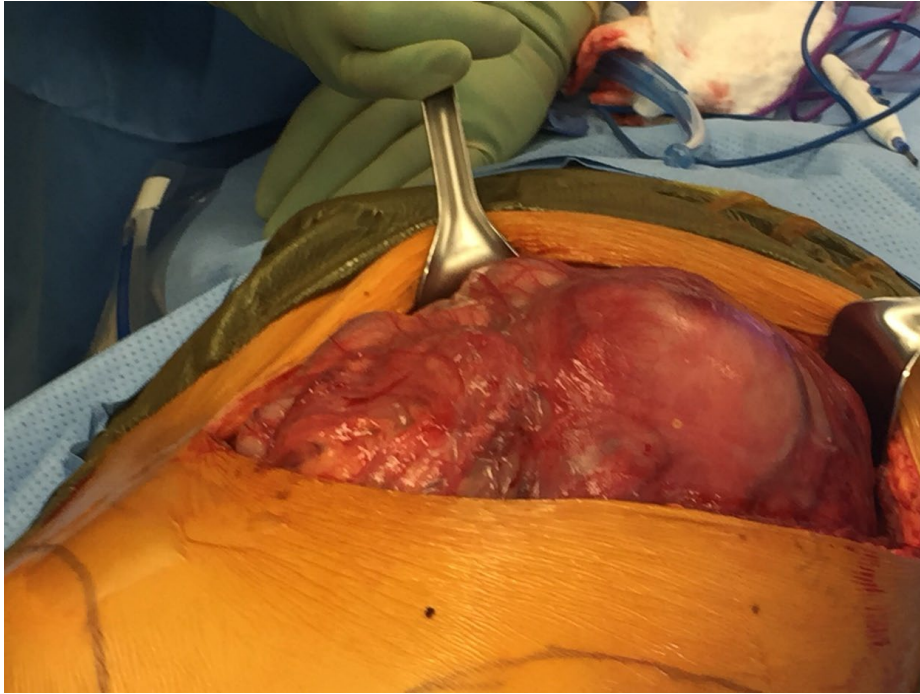




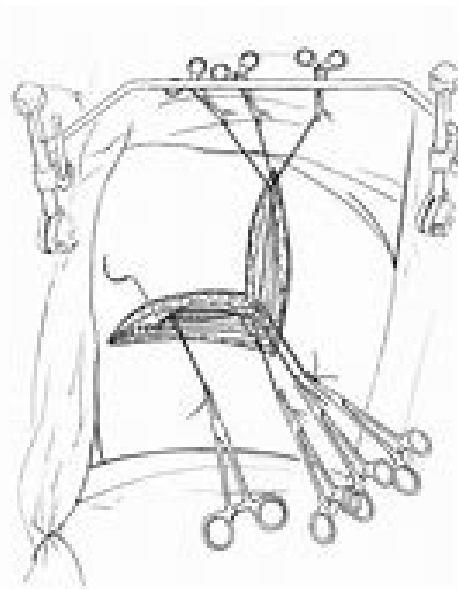
# Transverse Incisions

- Not much difference between horizontal vs. angled subcostal
- Advantages
  - Good exposure to lateral retroperitoneum
  - Can be extended to do bilateral cases
- Disadvantages
  - May not allow full exposure higher or lower
  - More limited in older kids as they get taller

# Transperitoneal Incisions



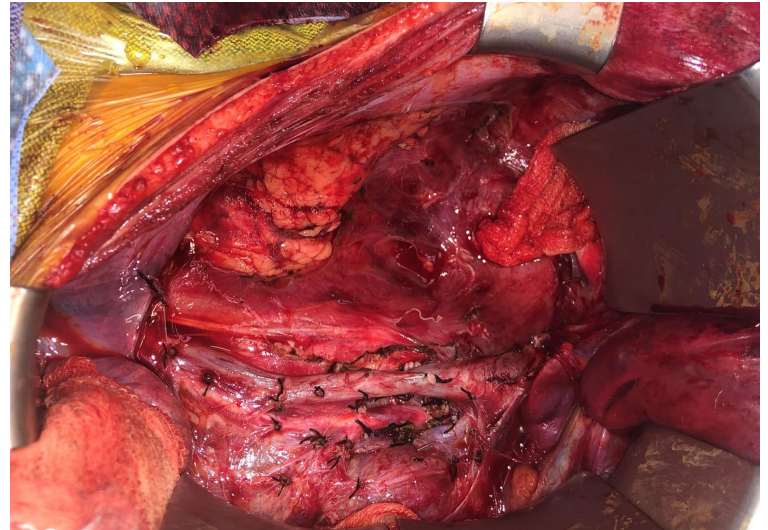
# Makuuchi Incision





# Makuuchi Incision

- Advantages
  - Great exposure to upper and lateral retroperitoneum
  - Especially good for older kids as they get taller and less wide
  - Good exposure for hepatic mobilization
  - Can be extended as a “Mercedes Benz” to do bilateral cases
  - Can combine with sternotomy for high level venous tumor thrombi
- Disadvantages
  - May not allow full exposure lower in pelvis
  - Cosmesis



# Makuuchi Incision

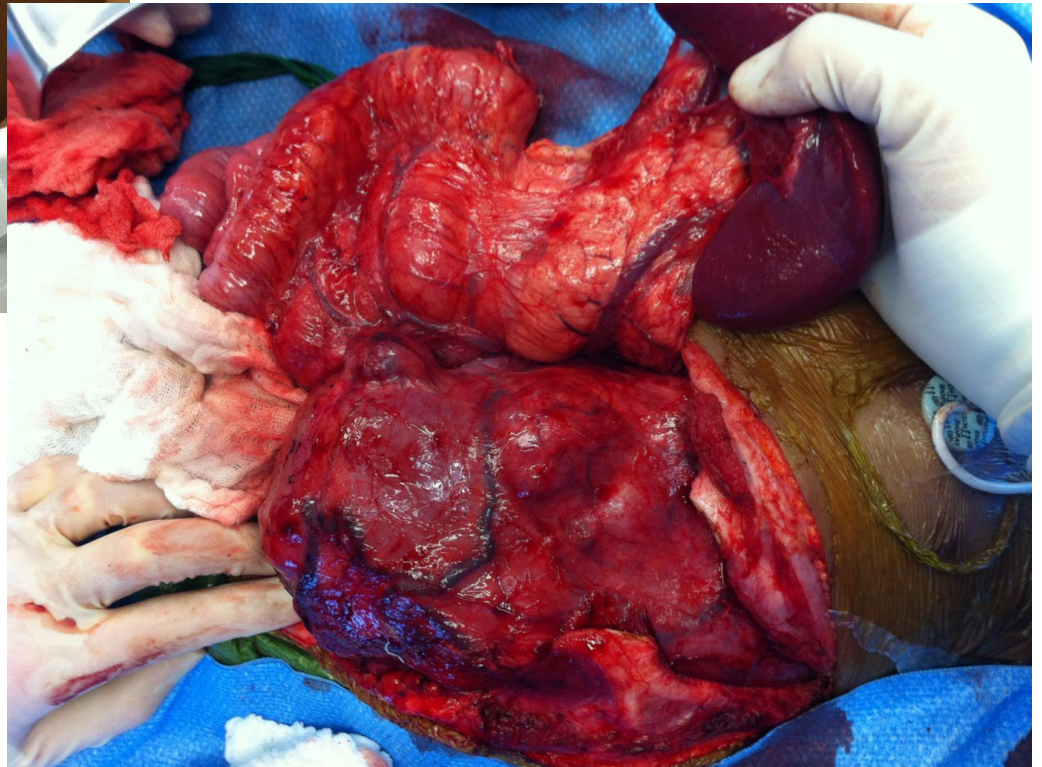


# Thoracoabdominal Incision

- Advantages
  - Maximal exposure to upper retroperitoneum
  - Can be extended to pelvis
  - Ideal for tumors that extend into retrocrural space
  - Can combined with ipsilateral pulmonary resections
  - Some like this approach for supra-hepatic venous tumor thrombi
- Disadvantages
  - Cannot easily access bilateral tumors
  - Violate thorax – require chest tube
  - Post op pain



# Thoracoabdominal Incision



# Intraoperative Considerations: Stage Properly

## Do

- Completely excise tumor and ureter to at least pelvic brim
- Remove small volumes of adjacent organs when needed
- Palpate ureter and renal vein intraoperatively
- Sample lymph nodes thoughtfully
- Maintain vascular control

## Do Not

- Take out large sections of organs
- Cut across or spill tumor
- Sample random lymph nodes
- Get too fancy





# Controversial Topics

## Image Based Feasibility of Renal Sparing Surgery for Very Low Risk Unilateral Wilms Tumors: A Report from the Children's Oncology Group

F. A. Ferrer,\* N. Rosen, K. Herbst, C. V. Fernandez, G. Khanna, J. S. Dome, E. Mullen, K. W. Gow, D. C. Barnhart, R. C. Shamberger, M. Ritchey and P. Ehrlich

From the Division of Pediatric Urology, Connecticut Children's Medical Center, Hartford, Connecticut



An enhanced recovery after surgery protocol in children who undergo nephrectomy for Wilms tumor safely shortens hospital stay

James K. Moon<sup>a,b</sup>, Rosa Hwang<sup>a</sup>, Frank M. Balis<sup>c</sup>, Peter Mattei<sup>a</sup>

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DOI: 10.1002/pbc.28212



ONCOLOGY: RESEARCH ARTICLE

## Minimally invasive surgery for unilateral Wilms tumors: Multicenter retrospective analysis of 50 transperitoneal laparoscopic total nephrectomies

Aurore Bouty<sup>1</sup> | Thomas Blanc<sup>2</sup> | Marc David Leclair<sup>3</sup> | Frederic Lavrand<sup>4</sup> | Alice Faure<sup>5</sup> | Aurelien Binet<sup>6</sup> | Julien Rod<sup>7</sup> | Mike O'Brien<sup>1</sup> | Sabine Sarnacki<sup>2</sup> | Michael Nightingale<sup>8</sup> | Yves Heloury<sup>1</sup> | Francois Varlet<sup>9</sup> | Aurelien Scalabre<sup>9</sup>

Journal of Pediatric Surgery (2013) 48, 1598–1603



Journal of Pediatric Surgery

www.elsevier.com/locate/jpedisurg

Review Articles

## Is adrenalectomy necessary during unilateral nephrectomy for Wilms Tumor? A report from the Children's Oncology Group ☆☆☆

Kathleen Kieran<sup>a,\*</sup>, James R. Anderson<sup>b</sup>, Jeffrey S. Dome<sup>c</sup>, Peter F. Ehrlich<sup>d</sup>, Michael L. Ritchey<sup>e</sup>, Robert C. Shamberger<sup>f</sup>, Elizabeth J. Perlman<sup>g</sup>, Daniel M. Green<sup>h</sup>, Andrew M. Davidoff<sup>a</sup>

# Why Staging Matters

- Tumor spill (56%), positive margins (47%), positive lymph nodes (25%) in Japanese series

**Table 3.** Impact of Surgical/Pathologic Factors on Event-Free Survival and Overall Survival in Patients With Stage III Favorable-Histology Wilms Tumor

Factor	Status	No.	4-Year EFS, % (95% CI)	<i>P</i> *	4-Year OS, % (95% CI)	<i>P</i> *
Nephrectomy	Up-front	419	89 (84.9 to 92.1)	.23	97 (95.4 to 99.1)	.22
	Delayed	116	85 (77.6 to 93)		95 (90.9 to 99.9)	
Lymph nodes	Negative	236	94 (90.2 to 97.4)	< .01	99 (97 to 100)	.09
	Positive	151	82 (74.9 to 89)		96 (92.2 to 99.6)	
	Unknown	148	84 (77.3 to 91.4)		95 (90.6 to 99)	
Gross residual disease†	Negative	394	88 (84.6 to 92)	.30	98 (95.9 to 99.4)	.051
	Positive	127	86 (78.6 to 93)		94 (88.9 to 98.7)	
LOH‡	Neither	377	91 (87.3 to 94.2)	< .01	97 (95.5 to 99.3)	.21
	16q only	96	83 (74.6 to 91.7)		97 (92.8 to 100)	
	1p only	56	75 (61.3 to 88.6)		92 (84.3 to 100)	
Peritoneal implants	Negative	364	86 (82.1 to 90.4)	.18	97 (94.3 to 98.7)	.34
	Positive	24	96 (87 to 100)		100	

Abbreviations: EFS, event-free survival; LOH, loss of heterozygosity; OS, overall survival.

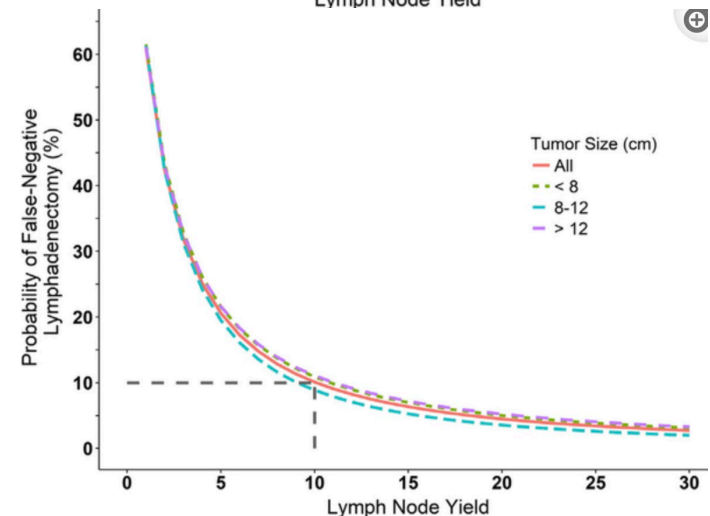
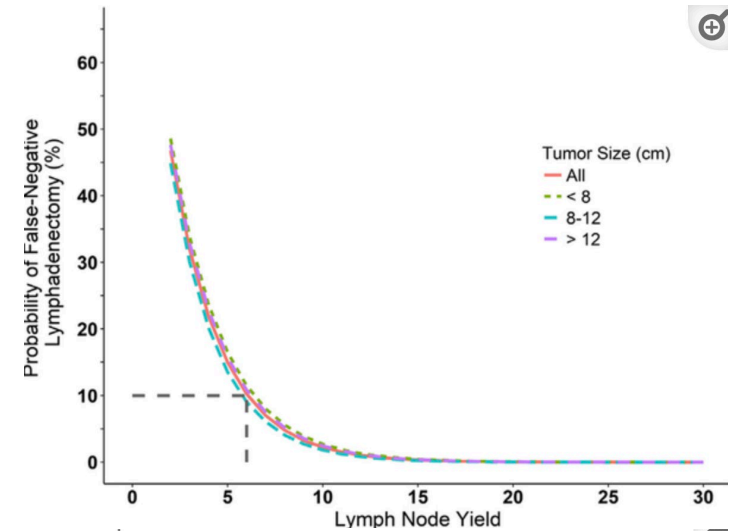
\*On the basis of the log-rank test.

†Includes all patients who had an up-front biopsy and then delayed nephrectomy.

‡Six patients with missing LOH data.

# Lymph Node Dissection

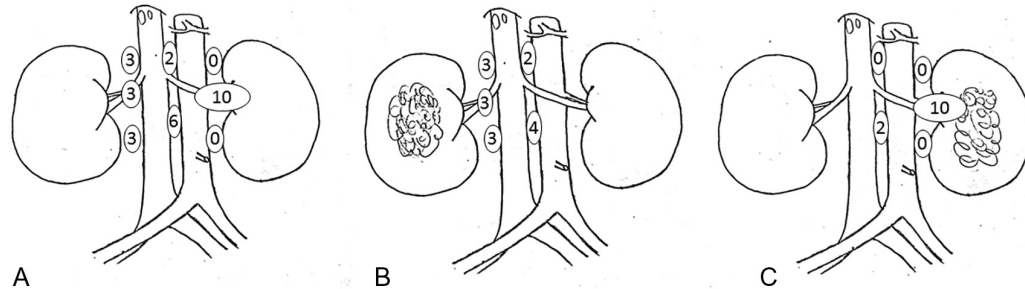
- Failure to sample lymph nodes (at all!) is most common protocol violation
  - 10-18% of surgeries
  - Some surgeons sample nodes that are not in renal drainage (e.g. mesenteric nodes)
- Nodal positivity may be more important for some subtypes of tumors
- Need 6-10 nodes to have “enough,” it seems



# Lymph Node Dissection

Standardizing lymph nodal sampling for Wilms tumor: A feasibility study with outcomes☆

Sajid S Qureshi <sup>a,b,\*</sup>, Monica Bhagat <sup>a,b</sup>, Mufaddal Kazi <sup>a,b</sup>, Seema A Kembhavi <sup>b,c</sup>, Subhash Yadav <sup>b,d</sup>,  
Badira C Parambil <sup>b,e</sup>, Vasundhara Smriti <sup>b,c</sup>, Akshay Baheti <sup>b,c</sup>, Maya Prasad <sup>b,e</sup>, Nehal Khanna <sup>b,f</sup>,  
Siddharth Laskar <sup>b,f</sup>, Tushar Vora <sup>b,e</sup>, Girish Chinnaswamy <sup>b,e</sup>, Nayana Amin <sup>b,g</sup>,  
Mukta Ramadwar <sup>b,d</sup>, Sanjay Talole <sup>b,h</sup>



Median 8 lymph nodes when templated dissection used.

Use of a lymph node sampling template was not associated with increased complications.

# Complications

- Bowel obstruction 5.1%
- Bleeding 1.9%
- Wound infection 1.9%
- Vascular injury 1.5%
  
- May be more accurate in institutional records
  
- Delays chemo (mean 6 days)
  
- Risk factors: tumor thrombus (OR=3.8), flank or paramedian incision (OR=2.0), tumor diameter >10 cm (OR=5.3), non-pediatric general surgeon (OR=9.0)\*

\*pediatric urologists had OR=0.7!

# In Summary

- Be meticulous
- Be thoughtful
- Be humble

# Thank You! Questions?

