



## Resection of intracardiac extension of Wilm's tumor with CPB– An experience with epidural analgesia

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### ABSTRACT

Wilm's tumor is the most common retroperitoneal tumor extending into the Inferior vena cava (IVC) in a child. In 1% of cases right atrial (RA) extension may be encountered. Anesthesia for intracardiac resection of tumor thrombus remains a challenge. We report 2 cases of Wilm's tumor, one extending upto IVC-RA junction and the other with RA involvement managed successfully with resection on cardiopulmonary bypass. In both patients epidural catheters were inserted for perioperative analgesia. Considerable controversy exists regarding insertion of epidural catheters in the face of systemic heparinization. However we report good outcomes in terms of analgesic efficacy, shorter perioperative course and no complications. A short discussion on the various methods of bypass employed for the removal of these tumors is included. We also discuss the utility of newer equipment like Transesophageal echocardiography (TEE) and ultrasound for perioperative anesthetic management.

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### Introduction

Wilm's tumor is the most common renal neoplasm in the pediatric population. 4-8% of tumors extend into the Inferior Vena Cava (IVC).<sup>1</sup> The management of these patients involves multispecialty approach with individualized surgical, oncological and anesthetic plans. We describe our experience in the management of 2 children presenting with IVC and RA extension of Wilm's tumor posted for radical nephrectomy with excision of tumor thrombus on cardiopulmonary bypass.

### Case Series

Written informed consent was obtained from parents of both children for publication.

The first child was a 23 month old girl weighing 10 kg. She presented with abdominal distension since 2 months. A CT scan of abdomen revealed a 11 X 8 X 8 cm solid heterogeneously enhancing mass in retroperitoneum arising from upper pole of right kidney extending into the Inferior vena cava (IVC). (Figure 1) Transthoracic Echocardiography revealed a hyperechoic mass extending upto the IVC-RA junction suggestive of a non obstructive tumor thrombus. There was no patent foramen ovale (PFO) and no tricuspid valve involvement. Biventricular function was normal. A diagnosis of right Wilm's tumour with IVC extension was made. Child was started on Tab Amlodipine 5 mg twice a day in view of hypertension which brought down the blood pressure to 110/90 mm Hg. 7 cycles of Adriamycin, Actinomycin and Vincristine chemotherapy was started following which there was a mild decrease in the size of the tumor.

The second child was a 5 year old boy weighing 13 kg who presented with abdominal distension since 3 months. CT scan was suggestive of a right Wilm's tumor extending upto the RA. Echocardiography showed a RA thrombus. There was no tricuspid valve involvement and biventricular function was normal. Child was given 3 cycles of neoadjuvant

chemotherapy with the same drugs which decreased the size of the abdominal tumor mass. However there was no decrease in the size of RA thrombus.



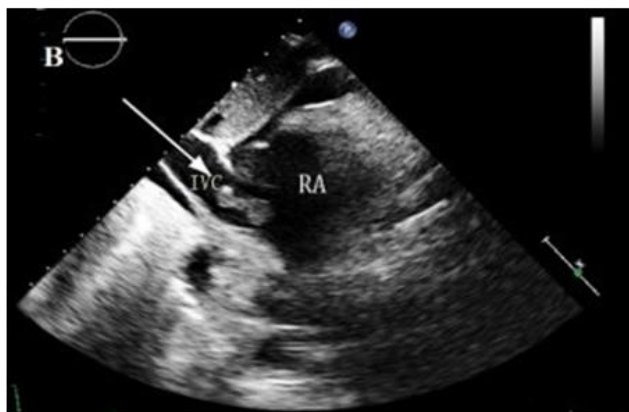
**Figure 1. CT scan showing large, right sided Wilm's tumor with IVC extension**

Preoperative investigations in both children revealed a normal coagulation profile with no derangement in renal or liver function.

A decision to perform a right radical nephroureterectomy with excision of tumor thrombus after establishing Cardiopulmonary bypass (CPB) in both children was made by a combined team of Pediatric surgeon, Oncologist, Cardiovascular surgeon and Paediatric Cardiac Anesthesiologist.

On the day of surgery both patients were premedicated with IV Midazolam 0.05 mg/kg and IV fentanyl 1 µg/kg. General anesthesia was induced with IV fentanyl 5 µg/kg, IV midazolam 0.5 mg/kg and muscle relaxation achieved with IV rocuronium 1 mg/kg. Following induction, right Internal Jugular Vein (IJV) and right femoral artery was cannulated

with ultrasound guidance. IV cefazolin 50 mg/kg and IV methylprednisolone 30 mg/kg was administered. 20 G epidural catheter was secured at T6-T7 interspace under all aseptic precaution and test dose of 0.1 ml/kg 1% adrenalized lignocaine was injected followed 20 minutes later by a bolus injection of 0.25% bupivacaine. Infusion of 0.25% bupivacaine was started for intra operative analgesia. The duration between epidural catheter placement and systemic heparinization was 1 hour 50 minutes in the first child and 2 hours 10 minutes in the second. Monitoring included electrocardiogram, invasive as well as non-invasive blood pressure, capnometry, central venous pressure, TEE (Transesophageal echocardiography), pulse oximetry, nasopharyngeal and rectal temperature, blood glucose, hematocrit, activated clotting time, arterial blood gas analysis, electrolytes, lactates and urine output. TEE immediately post induction reconfirmed absence of any inter-atrial communication and helped to visualize tumor extension (Figure 2) and central venous catheter tip. Anaesthesia was maintained on O<sub>2</sub>, air, Sevoflurane at 1 MAC with additional midazolam, fentanyl and vecuronium.



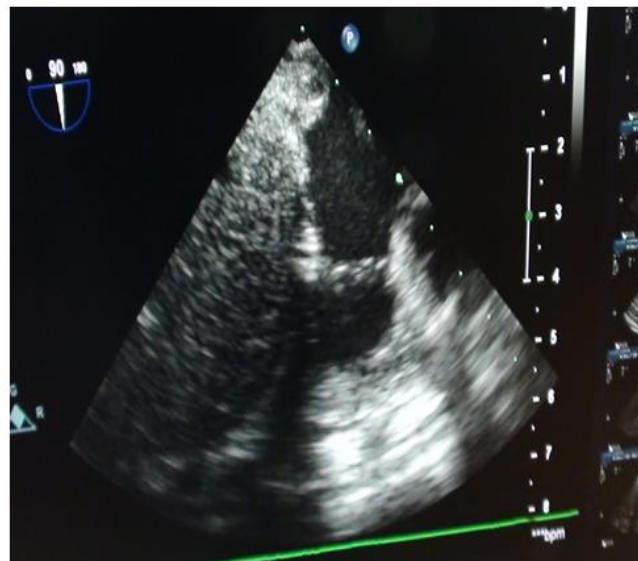
**Figure 2. TEE- midesophageal bicaval view showing thrombus at IVC-RA junction**

In the first child abdominal exploration with transverse incision was done. Right renal vein and artery was separated and ligated. Right radical nephrectomy with en bloc removal of tumor was performed. IVC was then slit open and thrombus removed upto suprahepatic IVC. Tumor thrombus above this level could not be removed. Midline sternotomy was then performed. Pericardium was opened and purse string sutures taken over aorta, SVC (superior vena cava) and abdominal IVC below renal vein opening. Systemic heparinization was done with 300 U/kg of heparin. Aorta, SVC and abdominal IVC was cannulated and CPB established. SVC was looped and snugged and abdominal IVC above cannula clamped. RA was then opened and tumour thrombus visualized on anterior wall of IVC-RA junction and removed en masse. Rest of RA was inspected for any tumour spread. Interatrial septum was found to be intact. RA was then closed. Patient was successfully weaned off CPB and heparin reversed with protamine. CPB time was 39 minutes. Child was shifted to ICU and ventilated postoperatively for 4 hours.

In the second child, a similar technique was employed for removal of RA thrombus. (Figure 3) CPB time was 41 minutes. Child was shifted to ICU and ventilated postoperatively for 3 hours 30 minutes.

Analgesia was maintained on epidural infusion of 0.125% bupivacaine and fentanyl (1µ/ml) till the 3rd post-operative day in both children. Wong-Baker six face pain rating scale was 3, 3 in the first and second post-operative day

respectively in the first child and 3,2 in the first and second postoperative day respectively in the second child. IV heparin 80U/kg 6 hourly was given on the first post-operative day in both children and level of anticoagulation monitored with aPTT. Recovery was uneventful. Histological examination of tumor mass was suggestive of Wilm's tumor with favourable prognosis in both cases. Both children were shifted to ward on the 4<sup>th</sup> post-operative day and subsequently discharged home.



**Figure 3. TEE showing RA thrombus**

#### Discussion

Wilm's tumor (nephroblastoma) is the most common renal neoplasm in the pediatric population accounting for 6% of all pediatric oncology cases. Intravascular extension is known with an incidence of 4-8% out of which intra atrial extension occurs in around 1% of patients.<sup>1</sup> This is more common on the right side due to shorter course of the right renal vein. Clinical presentation is commonly an asymptomatic abdominal mass which may sometimes be painful.<sup>2</sup> More than 50% of patients present with hypertension at the time of diagnosis due to a hyperreninemic state.<sup>2</sup> Coagulopathy may be present due to an acquired von Willebrand's disease.<sup>2</sup> Children may present with associated syndromes. These include Denys- Drash, WAGR (Wilm's tumor, aniridia, genitourinary anomalies, mental retardation), Beckwith – Widemann, Soto's syndrome and Trisomy 18. The results of US National Wilm's Tumor Study Group (NWTSG), French Société Internationale d' Oncologie Pédiatrique (SIOP), UK Children's Cancer Study group (UKCCSG) are used to stage, prognosticate and formulate treatment protocols. Diagnosis is usually established by ultrasound, computed tomography (CT) of abdomen and chest which influences clinical staging in case of lung metastasis. Intravascular extension places the patient in stage 3 and further echocardiographic evaluation is essential to document RA, tricuspid valve involvement, inter atrial communication and ventricular function. Intravascular involvement is further classified into three levels. Level 1- Infrahepatic, Level 2- Intrahepatic and Level 3- Suprahepatic or atrial extension.<sup>1</sup> Intravascular extension does not affect ultimate survival although risks like tricuspid valve obstruction, pulmonary embolism, tumor relapse or incomplete response to chemotherapy may be associated.<sup>1</sup> All 3 study groups (NWTSG, SIOP, CCSG) recommend aggressive pre nephrectomy chemotherapy to shrink the tumor size, treat concurrent metastasis in disseminated disease and reduce

surgical complication rate. Chemotherapy regimens usually include actinomycin D, doxorubicin and vincristine. Besides their propensity to cause immunosuppression, intractable nausea, vomiting and diarrhea, they have mainly hepatic metabolism and can cause fulminant hepatic failure. Vincristine can cause peripheral neuropathy, SIADH, convulsions, CNS depression. Doxorubicin can cause acute cardiomyopathy progressing to cardiac failure.<sup>2</sup>

In the presence of intracardiac extension, neoadjuvant chemotherapy is then followed by simultaneous intraabdominal and intracardiac resection after establishing partial CPB, mild hypothermic CPB without cardioplegia or CPB with deep hypothermic circulatory arrest (DHCA).

Anesthesiologist's role in caring for these patients arise during imaging procedures, CT guided biopsies, vascular access device (VAD) placement to facilitate chemotherapy administration and surgical resection of tumor. Pre Anesthetic check should elucidate history of any previous anesthetics and associated complications, known medical illness, syndromic features. A baseline blood pressure recording and all current medications including antihypertensives should be documented. Hemogram, coagulation profile with platelet function studies, serum creatinine levels, liver function tests should be noted and requisition made for adequate packed red blood cells, platelets, cryoprecipitate and fresh frozen plasma. ECG, echocardiography and holter monitoring is needed in children who have received doxorubicin cumulative dose >200 mg/m<sup>2</sup>.<sup>2</sup> Parents should be counseled regarding risks-benefit ratio of epidural analgesia, invasive hemodynamic monitoring, need for CPB and related complications, ventilatory support, and prolonged critical care requirement.

Intraoperative anesthetic management should focus on thermoregulation in infants and small children which may be further deranged with CPB or DHCA. Hypertensive or hypotensive crisis may occur. Intravenous access should be secured with a wide bore catheter in upper limb due to potential for severe hemorrhage. Central venous catheter placement should be done with ultrasound guidance due to the risk of dislodgement of tumor fragments and pulmonary embolism.

Epidural catheter placement in the face of systemic heparinisation is controversial. Spinal hematoma is rare but a potentially devastating complication. IVC compression due to the abdominal mass results in expansion of retroperitoneal venous collaterals which may make catheter placement more difficult. Trauma to epidural veins in presence of coagulopathy and dilated veins may result in large hematoma. Hence careful patient selection is required for catheter placement. There exists a reported risk of 1 hematoma for every 12,000 epidural catheterization.<sup>3</sup> For the safe use of epidural analgesia, the time from catheter placement to systemic heparinization should exceed 60 minutes and catheters should be removed when normal coagulation is restored. Patients should be closely monitored postoperatively for signs and symptoms of hematoma formation.<sup>4</sup> MRI is used for diagnosis and decompressive laminectomy should be performed within 6 hours in the event of a hematoma formation.

Epidural analgesia in both our patients helped in early weaning from mechanical ventilation, early mobilization, and good patient satisfaction with analgesia assessed by Wong Baker six face pain rating scale without any catheter related complications.

Transesophageal Echocardiography (TEE) helped us to rule out an inter atrial communication which can cause paradoxical embolus, to look for tricuspid valve occlusion, RA involvement, for correct placement of CVC tip, to guide placement of SVC- RA cannulae prior to establishing CPB, to guide fluid administration and to look for any residual thrombus post resection. (Table-1)<sup>5</sup>.

**Table 1. Important TEE views in intracardiac extension of Wilm's tumor<sup>5</sup>**

Structures visualized	TEE view
CVC tip placement	Midesophageal Bicaval
PFO	Midesophageal four chamber Midesophageal aortic valve short axis Midesophageal bicaval
Tricuspid valve	Midesophageal *RV inflow outflow Midesophageal four chamber Transgastric *RV inflow
SVC-RA cannulation	Midesophageal bicaval
Intracardiac tumor extension (RA)	Midesophageal bicaval Midesophageal aortic valve short axis

\*RV- Right Ventricle

CPB with or without hypothermia and circulatory arrest may achieve stable hemodynamic status during clamping of the IVC, thus providing a bloodless operative field for tumor resection and venous reconstruction. DHCA uses induced hypothermia (core temperature 15 to 18°) to protect the brain during cessation of blood flow. DHCA causes significant coagulation derangements and in the face of a pre-existing coagulopathy associated with Wilm's tumor adequate transfusion of platelets, FFP and cryoprecipitate is warranted for successful weaning from CPB. Due to the known complications of renal and hepatic failure, neurologic dysfunction, postoperative sepsis, and systemic coagulopathy associated with CPB, alternative techniques have been attempted.<sup>6</sup> VVB can be used in Level 1 and Level 2 intravascular thrombus extension to possibly reduce the risk of postoperative coagulopathy. It has the advantage that it does not require systemic anticoagulation, as the cannulas are precoated with heparin.<sup>7</sup>

Outcome of CPB in a single functioning kidney, pre-existing coagulopathy, neurological sequelae may alter perioperative outcomes in these patients. Repeated surgeries may be needed in these children due to metachronous tumors posing them at a risk for psychological disturbances and severely compromised renal function.

## Conclusion

The anesthetic management of Wilm's tumor with intracardiac extension remains a challenge. Management requires a multi-disciplinary approach by a pediatric surgeon, oncologist, cardiothoracic surgeon, pediatric cardiac anesthesiologist and Intensivist. Adequate preoperative optimization with parental counseling and individualized perioperative strategies with good analgesic techniques (epidural analgesia), judicious use of blood and blood products, decreased CPB time and use of newer equipment like TEE, ultrasound is the key to successful management.

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