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HYPERTROPHIC CARDIOMYOPATHY IN PREGNANCY: MATERNAL AND FETAL OUTCOMES AND CONSIDERATIONS.

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***Background*:** Women with severe Hypertrophic Cardiomyopathy (HCM) can successfully undergo pregnancy without significant maternal or fetal complications. The optimal management strategies however still remain fairly unknown.

Aims: The purpose of this study is to evaluate both maternal and fetal outcomes in women with HCM during pregnancy, labor and delivery and to clarify a management strategy that minimizes the adverse effects on both the mother and fetus.

***Methods*:** A database review of all women seen in the Department of Maternal/Fetal Cardiology from 1989–2005 was used to identify women with severe HCM defined as asymmetric septal hypertrophy ≥1.5cm by echo. Chart review was used to evaluate for progression of maternal disease as evidenced by cardiac deterioration (increased arrhythmias or hemodynamic compromise), maternal or fetal mortality, and fetal outcome. The management of these women during pregnancy, labor, and delivery was reviewed.

***Results*:** Thirteen women (16 pregnancies) with severe HCM were identified. Maternal echos were performed in all women at 20 and 30 weeks gestation and at 6 weeks postpartum. Fetal echos were obtained between 18–30 weeks gestation. Maternal echos demonstrated a median septal wall thickness of 2.3 cm (range 1.6–4.6 cm) and a mean predicted Doppler gradient in the left ventricular outflow tract of 37 mmHg (0–60 mmHg). There was no change on postpartum follow-up. Heart rate and/or rhythm control was achieved in most cases with Atenolol or Verapamil. One patient required intra-partum Esmolol. Holter monitoring revealed non-sustained ventricular tachycardia (VT) in 3/12 women. All women delivered in a perinatal special care unit with continuous monitoring of blood pressure and heart rate. CVP use was instituted early in the clinical experience however, over time was found to be unnecessary. All but one woman underwent assisted vaginal deliveries with a shortened 2nd stage of labor. There was one C-section. One infant of a diabetic mother had hypoglycemia. There were no other fetal or neonatal complications. Maternal complications included pulmonary edema, non-sustained VT and hypotension. There were no maternal or fetal deaths.

***Conclusions*:** Women with severe HCM can undergo successful pregnancy and delivery without significant complications or increased risk to the fetus. They are best managed in a high-risk center using a multidisciplinary team approach.

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OUTCOME OF CARDIAC THROMBI IN INFANTS.

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Cardiac masses in neonates are extremely rare and usually represent either tumors or thrombi. Management of cardiac tumors has been well described in this patient population, but a paucity of data exists on the management of cardiac thrombi, with the few reported cases focusing on outcomes following thrombolytic therapy (rt-PA, urokinase, or streptokinase). This study was undertaken to evaluate the outcome of cardiac thrombi in neonates who do not receive thrombolytic therapy.

Our echocardiographic database from November 1999 to November 2004 revealed 19 patients younger than 3 months of age [mean age = 41 ± 27 days (range 6 to 99 days)] when diagnosed with cardiac thrombi.

All 19 patients had a central venous line at the time of diagnosis. The median duration of central line prior to thrombus identification was 11 days (range 2 to 51 days). Sixteen (84%) had a thrombus attached to the endocardium [right atrium (n=11), right ventricle (n=2), left atrium (n=3)] and three (16%) had thrombi attached to the end of a central venous line within the right atrium. Nine patients (47%) had negative blood cultures, and complete resolution of the thrombus occurred in all of them within 24 ± 24 days (range 0 day to 77 days). These patients were treated with LMWH or heparin (n=2), antibiotics (n=2), both (n=2), surgical removal of the thrombus (n=2), or no therapy (n=1). In the 10 patients with positive blood cultures (53%), complete resolution occurred in 9 cases (90%) after a mean of 70 ± 58 days (range 5 to 166 days). Treatment consisted of either antimicrobials alone (n=5), or combination of LMWH or coumadin and antimicrobials (n=4). One patient doing well was lost to follow-up after 72 days. No patient had evidence of thrombus embolization.

The natural history of cardiac thrombi is that they resolve over time. Infected thrombi may require more prolonged therapy. Surgery is seldom required and thrombolytics are not usually necessary.

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DEALING WITH BEREAVEMENT IN THE NEONATAL INTENSIVE CARE UNIT (NICU).

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PURPOSE OF STUDY: To determine if post-mortem family conferences would have a positive impact on the healing process of families who experience the loss of their newborn. Through a questionnaire, we evaluated our experience and effectiveness in dealing with multiple aspects of the family's loss. We further evaluated our experience with the loss of a newborn to formulate a model in bereavement.

METHODS: We identified 370 newborns that expired in our NICU over an 11 year period (1994 – 2005). Under IRB approval, we sent a questionnaire to evaluate the family's emotional, social and vocational adaptation after the loss. Of the 370 families identified, 104 were excluded due to no forwarding address and/or incomplete database. Of the remaining 266 families, 29 (11%) responded.

RESULTS (N = 29)

GA (wks) (23-41)	Time of Death (days) (1-160)	Maternal Age (years) (22-42)	Post-Loss Divorce	Employed Prior/After Loss	Children After Loss
28.69 ± 5.73	20.03 ± 37.34	31.9 ± 5.73	1 (3%)	22 (76%)	13 (45%)

CONCLUSIONS: Based on our data, a specific bereavement team should be identified to implement a consistent approach and follow up for families that experience the loss of a newborn. A post mortem conference appears to reduce the intensity of the grieving process and provide comprehensive support. Offering this conference should be mandatory. Autopsy consent and obstetrical involvement can be improved. This approach can provide experience and education in this humanistic side of medicine relevant to all members of the healthcare team who care for critically ill newborns.

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REDUCED NEONATAL CANDIDA INFECTION AS A RESULT OF MEDICAL PRACTICE INTERVENTION

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Objective: To assess the impact of a medical practice intervention on the incidence of *Candida* spp. sepsis among neonates with a birth weight of ≤ 750 grams admitted to a level 3 neonatal intensive care unit (NICU).

Methods: In July 1996 implementation of medical practice included;(1) administration of oral nystatin (every 6 hours for the first week of life and with each antibiotic course during the first four weeks of life). (2) Modifying parameters for extubation, (3) Early discontinuation of central lines (UAC, UVC, PCVC, CL), (4) Parenteral nutrition (PN) and antibiotics. We reviewed charts and collected data from January 1, 1995 to December 31, 1998. Neonates who died at ≤ 3days of age or were admitted at ≥ 7days of age were excluded. The intervention group consisted of 69 neonates, admitted between July 1, 1996 and December 31, 1998 and the control group consisted of 45 neonates, admitted between January 1, 1995 and June 30, 1996. We measured the outcome by using student t test and standard logistic regression techniques. The above management plan was continued after the study period. Retrospective analysis of the incidence of Candida infection in NICU was followed for 1999 to 2004.

Results: The intervention group experienced fewer episodes of *Candida* spp. sepsis, OR 0.07 (p=<0.0002) adjusting for gestational age, mode of delivery and number of days of central vascular access. The estimated gestational age was longer (EGA) (24.9 vs. 24.4 p=<0.05) and more neonates received feedings (93% vs. 73%, p=<0.05). Within the intervention group, exposure to high humidity environment was associated with a higher risk of *Candida* spp. sepsis, OR 10.5 (p<0.06). The incidence of candida spp. During 1999 to 2004 remained between 0% to 3%.

Conclusion: Oral nystatin administration substantially reduced the risk of *Candida* spp. sepsis among neonates with birth weight ≤ 750g, while vaginal delivery was an independent risk factor. High humidity environment may be an additional risk factor for developing *Candida* spp. sepsis.

	Before	intervention	After					
Period	1/1/95-6/30/96	7/1/96-12/31/98	1999	2000	2001	2002	2003	2004
Incidence	36%	6%	1%	2%	2%	3%	1%	0%

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ADVERSE EVENTS IN PEDIATRIC KETAMINE SEDATIONS WITH AND WITHOUT MORPHINE PRETREATMENT

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Purpose: To assess outcomes between two groups of patients receiving ketamine for procedural sedations in our pediatric emergency department. Our hypothesis is that there is no difference in the number of adverse events in ketamine sedations with and without morphine pretreatment.

Method: This was a retrospective cohort study of all ketamine sedation records over 15 months. The number and types of adverse events between patients with and without morphine pretreatment were compared, using a z-score and p-values were obtained. The possible influence of midazolam co-administration was examined, using Fisher's exact and Pearson chi-square tests.

Results: 858 sedations were reviewed. Age, weight, and medication dosages were similar in each group. 21 adverse events were recorded in the group of patients with no morphine pretreatment; and, 13 adverse events in the group with morphine pretreatment. No significant differences were found for the number or types of events. There was no difference for the frequency of midazolam co-administration, Pearson chi-square, p = 0.994; nor for the number of adverse events in each group, Fisher's exact test, p = 0.465. The mean time from morphine administration to procedural sedation was 114.7 minutes. One adverse event occurred in the 15 min or less time interval.

Conclusions: We found no increased adverse events with morphine pretreatment in ketamine sedations for children. Prospective studies to validate these findings, including an effect of timing of analgesia administration are warranted.

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EARLY RECURRENCE OF CRESENTRIC GLOMERULONEPHRITIS AFTER PEDIATRIC RENAL TRANSPLANTATION:

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Introduction: Recurrence of primary disease such as focal segmental glomerulosclerosis or hemolytic uremic syndrome may cause early graft dysfunction after pediatric kidney transplantation. We report the unusual occurrence of early graft dysfunction following kidney transplant in two pediatric cases due to biopsy proven recurrence of cresentric glomerulonephritis (CGN) which is very rare.

Methods: Of the 75 renal transplants performed in patients aged <18 years at our center from 1996–2004, 2 patients had an early recurrence of CGN (in less than 7 days) leading to acute graft dysfunction. In order to rule out the different etiologies which could present as CGN in the biopsy, tests for anti-glomerular basement membrane antibodies (anti-GBM), anti-nuclear antibodies (ANA), anti-neutrophil cytoplasmic antibodies (ANCA, p-perinuclear staining, c-cytoplasmic granular staining), serum complement C3, C4 levels and immunofluorescent (IF) tests were done.

Results: Patient A was able to recover her renal function following aggressive treatment with plasmapheresis, intravenous (IV) cyclophosphamide and methylprednisolone. Twelve years after the transplant, her serum creatinine continues to be near normal (1.5 mg/dl). In patient B, plasmapheresis before and after the transplantation along with IV cyclophosphamide and methylprednisolone did not prevent graft failure, who is currently on chronic peritoneal dialysis.

Conclusion: Early recurrence of CGN though uncommon, should be considered in the differential diagnoses of early graft dysfunction after renal transplantation in children. Early diagnosis and treatment in some patients with CGN can lead to recovery of graft function thus improving the positive outcome after transplantation.

Tests	Anti-GBM	ANA	ANCA	Serum C3,C4	Biopsy (IF)
Patient A	(-)	(-)	p-ANCA (-) c-ANCA (-)	Normal	IgG IgA IgM C3 (-) (+) (+) (+)
Patient B	(-)	(-)	p-ANCA (+) c-ANCA (-)	Normal	IgG IgA IgM C3 (-) (-) (-) (-)