

SURVEY OF CURRENT CANADIAN PRACTICE IN THE MANAGEMENT OF NEWBORNS WITH HEMOPHILIA



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INTRODUCTION

- Newborns with hemophilia A and B are at increased risk of intracranial hemorrhage^{1,2}
- Some aspects of postnatal care – neuroimaging, empiric use of factor concentrate, administration of vitamin K – may contribute to the diagnosis and prevention of intracranial hemorrhage
- Only one published guideline³ addresses these issues, and it is unknown how well current practice matches this guideline
- There are no data comparing the practices of hematologists and neonatologists

OBJECTIVES

- To describe current management of newborns with hemophilia in Canada – with respect to timing of hematology consultation, vitamin K administration, neuroimaging, and empiric treatment with hemostatic products – in specific clinical scenarios
- To compare the practices of hematologists and neonatologists/pediatricians
- To identify areas of consensus that might be used as the basis for clinical practice
- To identify areas that lack consensus as important research priorities

METHODS

- Electronic and paper survey instrument sent to hematologists and neonatologists/pediatricians practicing in Canada, identified from membership lists of professional groups
- Survey conducted in August / September 2010
- Response proportions between groups compared using 2-sided Pearson chi-square or Fisher's exact tests

RESULTS

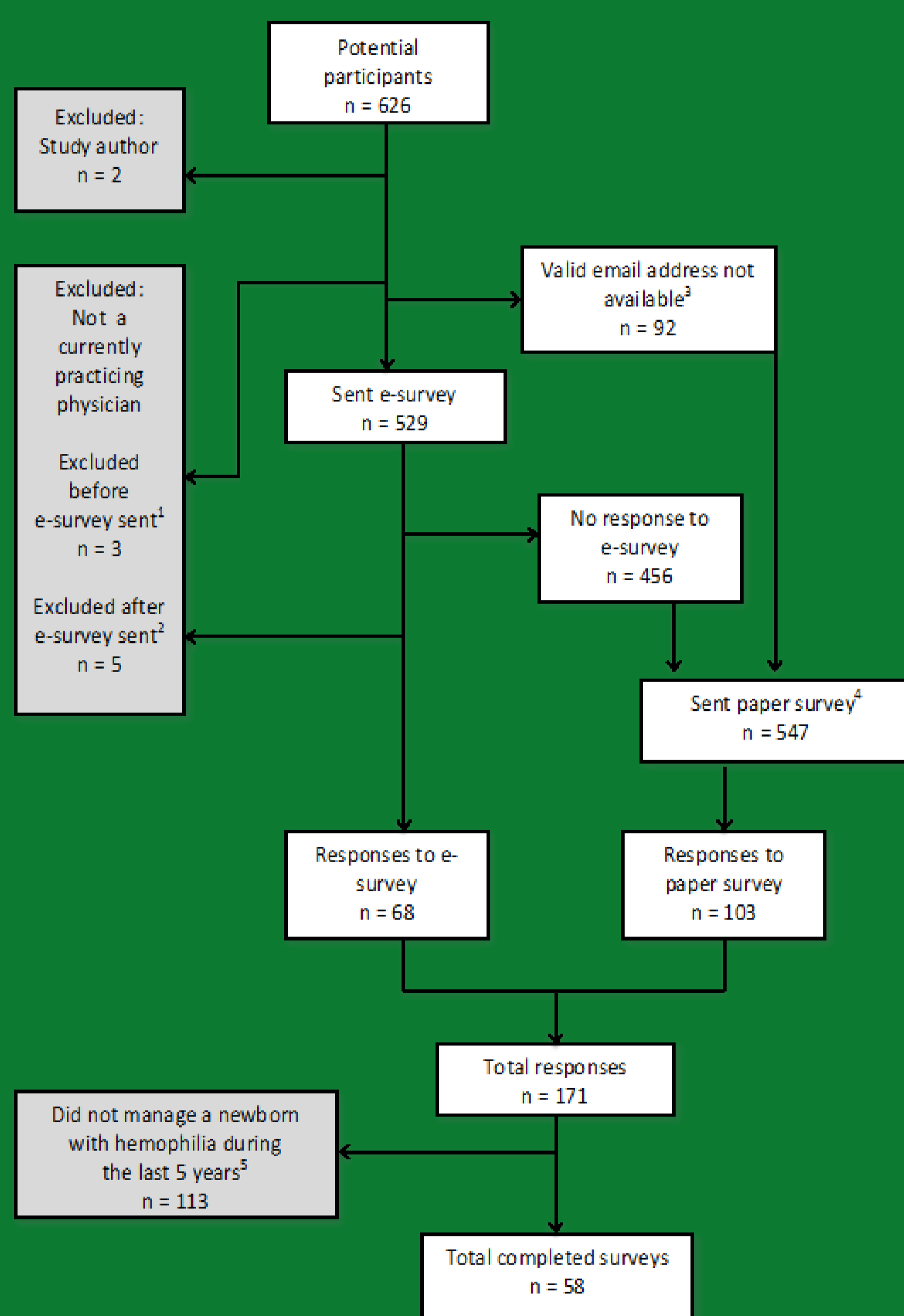


Figure 1: Conduct of the Survey

¹ Physicians, not currently practicing: retired (n = 1) and maternity leave (n = 2). ² Non-physicians identified by internet research: registered nurse (n = 1), graduate student (n = 1), researcher (n = 3). ³ Valid email address not available: Canadian Hematology Society members (n = 49), bounced email (n = 32), no email address available (n = 6), previously opted out of Survey Monkey (n = 5) ⁴ A mailing address was not available for one person who had been sent the e-survey. ⁵ Six respondents indicated by email that they do not treat pediatric patients with hemophilia; this was considered equivalent to having responded to the survey and indicating that they had no recent experience managing newborns with hemophilia

Scenario	Description
Scenario 1: Maternal Carrier, Uncomplicated Delivery, Well Newborn	A 26-year-old G2P1 woman gives birth to a boy at 39 weeks' gestation by uncomplicated vaginal delivery. The mother is a known carrier of a mutation for severe hemophilia A. The baby boy is clinically well.
Scenario 2: Maternal Carrier, Difficult Delivery / Caesarian Section, Well Newborn	A 29-year-old G1P0 woman gives birth to a baby boy at 39 weeks' gestation. The mother is a known carrier of a mutation for severe hemophilia A. Vaginal delivery is attempted, but is converted to an emergency caesarian section because of fetal bradycardia and difficulty extracting the baby's head. The baby boy is clinically well after birth, but he has facial bruising and a cephalohematoma.
Scenario 3: Prenatal Diagnosis, Uncomplicated Delivery, Symptomatic Newborn	A 33-year-old G3P2 woman gives birth to a baby boy at 38 weeks' gestation. Prenatal testing has confirmed that the baby has a factor VIII mutation associated with severe hemophilia A. The birth is by uncomplicated vaginal delivery. Within 6 hours of birth, the baby is observed to feed poorly, and subsequently the baby has a seizure.

Box 1: Clinical Scenarios Used in the Survey Instrument

	Consult Before Delivery (%)		Consult at Delivery (%)		Consult at Diagnosis (%)		p - value
	Heme	Neo/Peds	Heme	Neo/Peds	Heme	Neo/Peds	
Scenario 1	90	62	7	3	3	35	0.01
Scenario 2	86	59	14	7	0	34	0.003
Scenario 3	97	89	3	7	0	4	0.476

Table 1: Response Proportions for Timing of Hematology Consultation

Heme, Hematologists. Neo/Peds, Neonatologists/Pediatricians

DISCUSSION

- Few areas of consensus:
 - No empiric factor for well newborns after uncomplicated delivery
 - Cranial ultrasound for bruised asymptomatic newborns after difficult delivery
- Important questions without consensus:
 - Preferred route of vitamin K administration?
 - Empiric factor concentrate for bruised asymptomatic newborn after difficult delivery?
 - Need for neuroimaging of asymptomatic newborns after uncomplicated delivery?

- Systematic differences between hematologists and neonatologists / pediatricians:
 - Timing of hematology consultation for care of the fetus / newborn
 - Use of oral vitamin K for newborns with prenatal diagnosis
 - Use of specific factor concentrate for symptomatic newborns

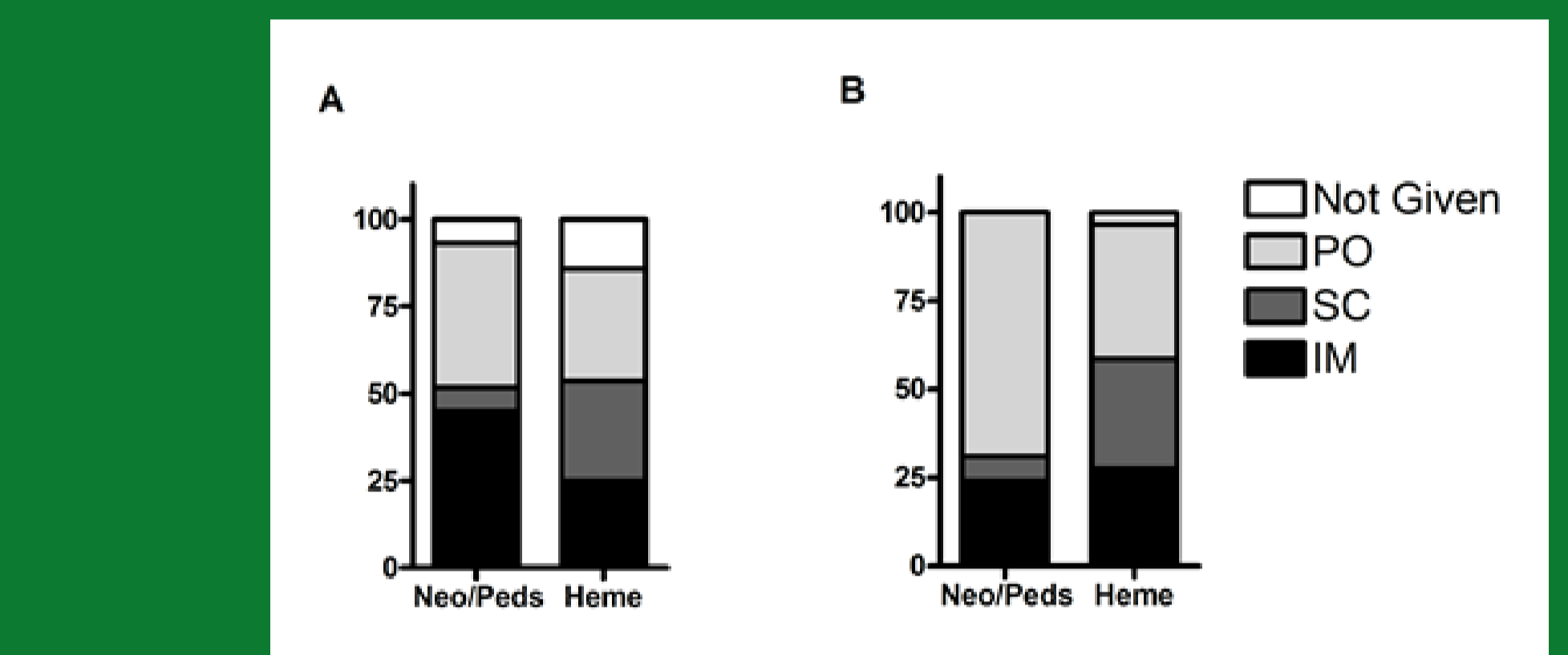


Figure 2: Response Proportions for Questions Involving Vitamin K Administration

A: Scenario 1. p > 0.05. B: Scenario 3. p = 0.043. Neo/Peds, Neonatologists/Pediatricians. Heme, Hematologists. PO, per os. SC, subcutaneous. IM, intramuscular.

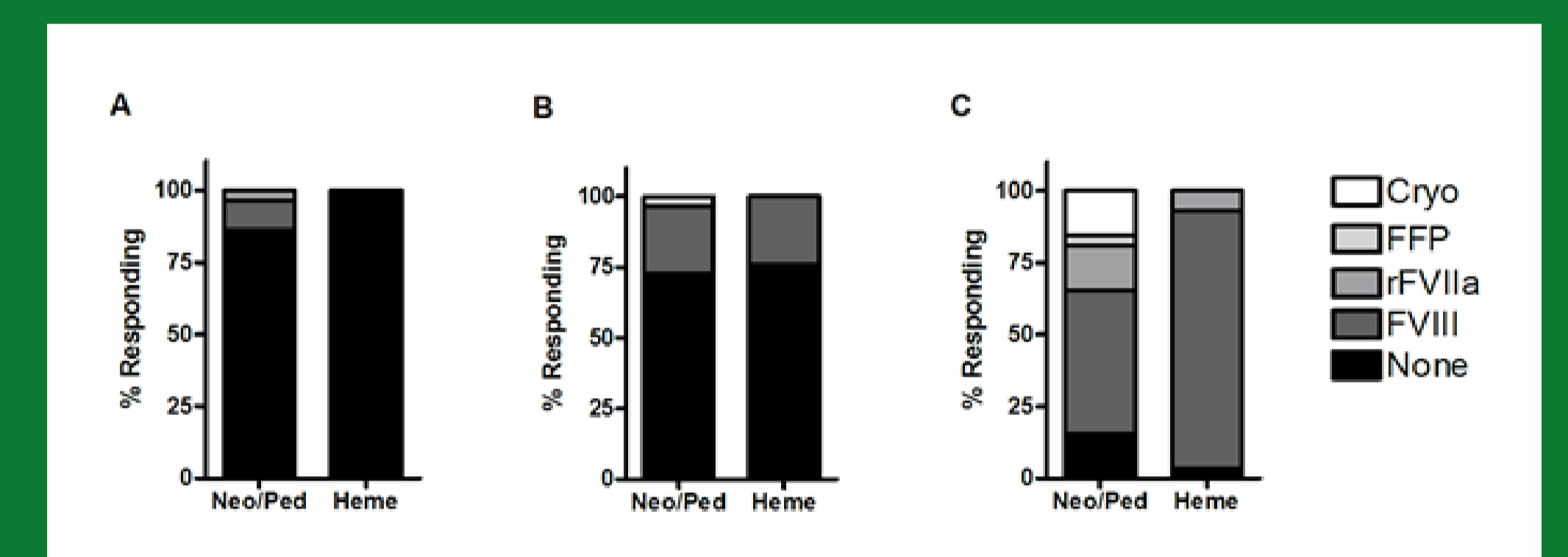


Figure 3: Response Proportions for Questions Involving Empiric Treatment

A: Scenario 1. p > 0.05. B: Scenario 2. p > 0.05. C: Scenario 3. p = 0.02 Neo/Peds, Neonatologists/Pediatricians. Heme, Hematologists. Cryo, cryoprecipitate. FFP, fresh frozen plasma. rFVIIa, recombinant activated factor VII. FVIII, factor VIII (either plasma-derived or recombinant).

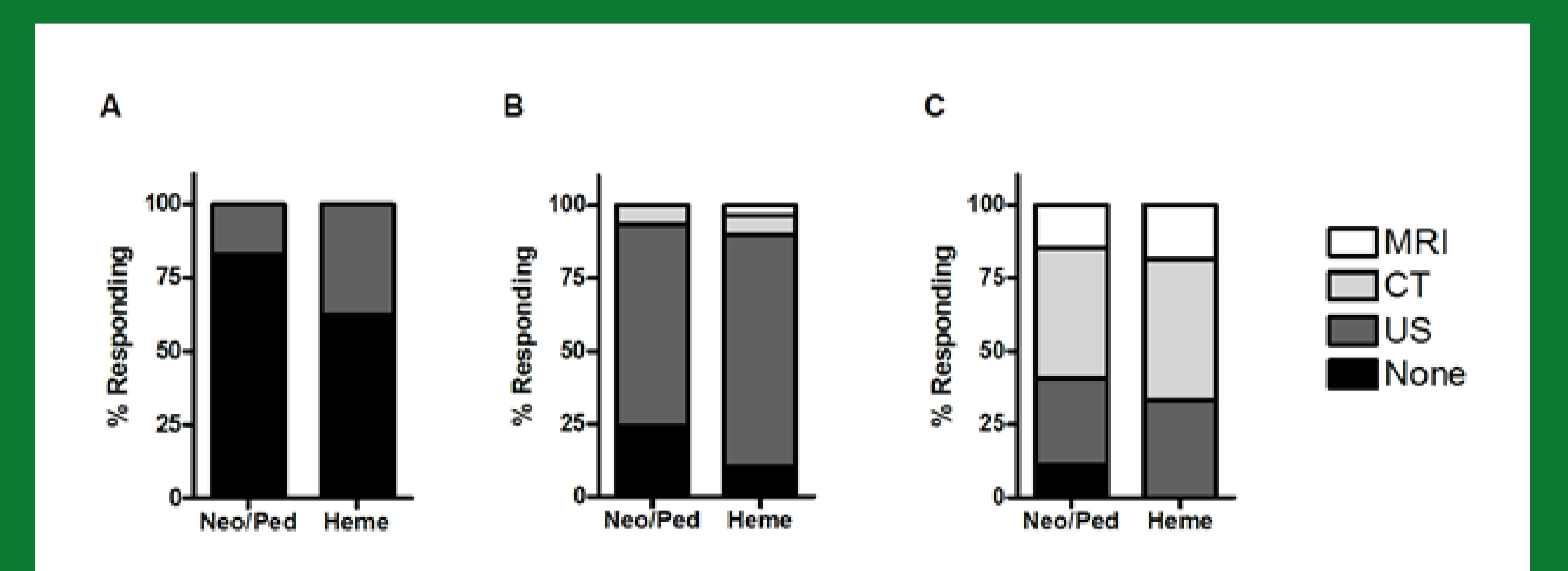


Figure 4: Response Proportions for Questions Involving Cranial Imaging

A: Scenario 1. p > 0.05. B: Scenario 2. p > 0.05. C: Scenario 3. p > 0.05. Neo/Peds, Neonatologists/Pediatricians. Heme, Hematologists. US, ultrasound. CT, computed tomography. MRI, magnetic resonance imaging.

CONCLUSIONS

- Need for improved communication between hematologists and neonatologists
- Prenatal consultation of pediatric hematology services
- Guidelines to standardize care should be developed and adopted
- Prioritize research to identify strategies for optimal care of newborns with hemophilia

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