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ORIGINAL ARTICLE

The impact of preparation and support procedures for children with sickle cell disease undergoing MRI

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Abstract
Background Children with sickle cell disease (SCD) often undergo MRI studies to assess brain injury or to quantify hepatic iron. MRI requires the child to lie motionless for 30–60 min, thus sedation/anesthesia might be used to facilitate successful completion of exams, but this poses additional risks for SCD patients. To improve children's ability to cope with MRI examinations and avoid sedation, our institution established preparation and support procedures (PSP).
Objective To investigate the impact of PSP in reducing the need for sedation during MRI exams among children with SCD.
Materials and methods Data on successful completion of MRI testing were compared among 5- to 12-year-olds who underwent brain MRI or liver R2*MRI with or without receiving PSP.
Results Seventy-one children with SCD (median age 9.85 years, range 5.57–12.99 years) underwent a brain MRI (n=60) or liver R2*MRI (n=11). Children who received PSP were more likely to complete an interpretable MRI exam than those who did not (30 of 33; 91% vs. 27 of 38; 71%, unadjusted OR=4.1 (P=0.04) and OR=8.5 (P<0.01) when adjusting for age.
Conclusion PSP can help young children with SCD complete clinically interpretable, nonsedated MRI exams, avoiding the risks of sedation/anesthesia.

Keywords Sickle cell anemia · Sedation · MRI · MRA · Child life specialist · Preparation and support procedure · Children

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Introduction

Children with sickle cell disease (SCD) are at an increased risk of developing ischemic brain injury, including overt strokes and silent infarcts [1, 2]. The frequency of primary overt stroke in homozygous HbSS children is 5–10% [3], with cumulative incidences of primary stroke of 11% by age 20 years [1, 4]. Chronic blood transfusion therapy is considered the standard of care in children with SCD who are known to be at high risk for stroke [5, 6]. Children with SCD often undergo imaging studies to detect or monitor the neurological complications of their disease [7] as well as iron overload [8], a common complication of repeated blood transfusions. MRI and magnetic resonance angiography (MRA) are among the most frequently used diagnostic tests to evaluate the brain for evidence of new disease or progression of existing disease [7, 9, 10] and R2*MRI has been

shown to accurately quantify iron deposition in the liver [11, 12].

To obtain accurate information and reduce motion artifact during MRI testing, children are required to remain motionless during the exam. Avoiding movement during an MRI test can be particularly challenging for young children, especially considering the long duration of many MRI exams (30–60 min). Many hospitals use sedation/anesthesia to ensure stillness and successful completion of the MRI examination. In children with SCD, however, there are increased risks involved with the use of sedation/anesthesia. Sedation can increase vaso-occlusive complications from SCD including pain and acute chest syndrome [13–15]. To avoid these complications, children receive aggressive intravenous hydration before undergoing sedation/anesthesia. In some cases, blood transfusions are also required. These preventive measures can effectively reduce risk but are burdensome to children and families and increase the cost of the diagnostic procedure significantly. Furthermore, avoiding sedation in a young child can allow performance of certain MRI exams that cannot be completed under sedation, such as functional MRI techniques.

With the goal of reducing the risks and high costs associated with sedation/anesthesia, a program was implemented at St. Jude Children's Research Hospital to deliver preparation and support procedures (PSP) for young patients (5–12 years old) with SCD undergoing MRI testing of either the brain or liver. The PSP program aims to educate and prepare children with SCD and their families about an upcoming MRI procedure, with the goal of improving cooperation and coping with MRI testing while reducing the need for sedation/anesthesia and the subsequent risks associated with it. In this retrospective study, we reviewed whether use of the PSP program affected the ability of young children to successfully complete brain MRI or liver R2*MRI exams without the use of sedation/anesthesia.

Materials and methods

Study participants

This retrospective study was approved by the Institutional Review Board at St. Jude. Data were reviewed from the medical records of all children with SCD (of any genotype) at our institution between the ages of 5 and 12 years who underwent a conventional MRI of the brain (with or without MRA) or an R2*MRI of the liver (for iron quantitation) between September 2008 and November 2009. R2*MRI of the liver was included in this analysis because this exam is increasingly used in children with SCD and iron overload and requires patient cooperation with the breath-holding technique. Demographic and clinical data were collected. Children younger than 5 or 13 years or older were excluded because children who are ≤5 years of age usually require sedation/anesthesia for MRI procedures, and those ≥13 years of age are able to cope reasonably well with MRI testing without sedation/anesthesia or a support procedure.

PSP program

Recognizing the need for a better means to prepare children with SCD for diagnostic imaging testing without using sedation/anesthesia, the Child Life Program at St. Jude offered PSP to children with SCD who were scheduled to undergo MRI exams between September 2008 and November 2009. The objectives of the PSP program were to reduce test-associated anxiety, recognize patient-specific obstacles to performing the diagnostic test, identify each child's individual coping strategies, and help children develop a sense of control. The ultimate goal was to improve coping and cooperation with MRI testing in order to obtain an adequate and clinically interpretable MRI exam without using sedation/anesthesia.

With this goal in mind, the unique background of a child life specialist is ideal in this setting. Child life specialists are trained professionals with degrees in human growth, child development and related areas. To become certified, child life specialists must complete a supervised clinical internship, pass a certification examination administered by the Child Life Council and follow guidelines for continuing professional development throughout their careers [16]. Because of this specialized training, child life specialists are able to focus on children's developmental and psychosocial needs within the hospital setting and promote effective coping for children facing challenging or stressful experiences.

Children were referred to the PSP program by their treating clinicians, who made these decisions during the children's regularly scheduled clinic visits. A referral was made on the basis of caregiver's availability to meet with the child life specialist. Only stable patients were referred to the PSP program and acutely ill patients were referred for sedation/anesthesia because of the urgent need of their MRI exams. All children with SCD participating in the PSP program were evaluated by a certified child life specialist within 30 days of MRI testing. During this meeting, the MRI procedure was explained to the child and his or her caregivers, and the child's prior experience (anxiety level, use of sedation, etc.) with MRI testing was reviewed (Table 1). The PSP program was provided to children by only one child life specialist. The child life specialist used a small model MRI machine, pictures of the MRI suite and recordings of MRI sounds to prepare the child for the procedure. The sequence of events for the MRI exam and sensory information, including sight, sound and touch, was provided using age-appropriate explanations. Children and caregivers had an opportunity to ask questions. Each child

Table 1 Preparation and support procedure steps

Prior to MRI testing
Preparation
• Child life specialist meets with family and child
• Teaching materials shown (model MRI machine, pictures of MRI suite and MRI sounds)
• Sequence of events explained
• Sensory information explained
Rehearsal
• Child empowered with a job to do during MRI exam
• Child rehearses the job
• Coping plan developed in conjunction with child and parent
During MRI testing
Support
• Presence of parent or child life specialist in MRI suite
• Touching of child's hands or feet for reassurance
• Squeeze ball in hand if needed
• Updating child on MRI progress
• Visual or audio distraction

was given a specific job, such as holding still during a brain MRI, or holding his or her breath for 10-s intervals during a liver R2*MRI. Next, the child life specialist led the child in rehearsing his or her job, while giving the child time to ask questions and develop a coping plan for the procedure. At the end of this session, an individualized plan was developed to address the specific aspects of the MRI testing. The coping plans were tailored to the individual needs of each child and family and included options such as the presence of one caregiver and/or the child life specialist in the room during the MRI exam; holding onto a squeeze ball in case the child felt the urge to move; touching of the child's hand or feet by the parent or the child life specialist during the procedure to remind the child of the presence of a support person in the exam room; updating the child on the progress of the MRI testing (how much time was left, etc.); and watching a movie (an option only during a brain MRI), listening to music or listening to a book on tape.

The costs associated with implementing the PSP program were relatively low. The tabletop model MRI machine was purchased for less than US $600 and included an audio recording of MRI sounds and a doll to fit the model MRI machine. Other supplies included a digital camera and printer paper to create the photo books, as well as squeeze balls, which cost less than $200 altogether. Audio distraction files were downloaded from free websites or purchased audio books. The most significant cost to implementing the PSP program was the MRI-compatible movie system (ranging from $4,000 to $7,000), which includes a projector with special lens, a screen, cables and mirrors that attach to the MRI head coil.

Statistical analyses

An MRI test was defined as successful when it yielded interpretable results (by a neuroradiologist or pediatric radiologist) and did not require the use of sedation/anesthesia. Median ages were compared between patient groups by the Mann-Whitney-Wilcoxon test. Associations between gender and prior MRI exposure were compared using the chi-square test. Logistic regression models were used to evaluate the association between PSP and completion of an interpretable MRI exam, with and without controlling for age. Model-based odds ratio estimates are presented with 95% confidence intervals. *P*-values less than 0.05 were considered statistically significant. All data were analyzed using SAS version 9.2 (Cary, NC).

Results

A total of 87 children with SCD underwent MRI testing (brain MRI or R2*MRI of the liver) from September 2008 to November 2009. However, 16 (18.4%) children were prescribed sedation prior to being considered for PSP and were therefore excluded from the main outcome analysis. The decision to prescribe sedation/anesthesia in these 16 cases was entirely at the discretion of the treating clinician and the reasons for this choice were not always available, but in most cases there was an emergent need for the MRI testing. The median age of the remaining 71 children was 9.9 years (range 5.6–12.9 years). Of these children, 65 (91.5%) had HbSS, 5 (7.0%) had HbSβ^0 thalassemia, and 1 (1.4%) had HbSC genotypes. Sixty (84.5%) children underwent brain MRI and 11 (15.5%) underwent a liver R2*MRI.

The child life specialist offered PSP to 33 (46.5%) children; among the specific options for activities, 7 chose to listen to music, 25 chose to watch a movie, and 1 chose to listen to an audio book. There were no differences in gender among children who received PSP and those who did not; however, PSP participants were younger than non-participants (median ages 8.9 vs. 10.9 years, $P=0.0002$). There were no significant differences in prior exposure to MRI among PSP and non-PSP participants (Table 2). The age among PSP participants who failed an MRI exam ranged from 5.4 to 7.3 years. The success rates in coping and completing an interpretable MRI test did not differ significantly between children undergoing liver R2*MRI and those undergoing brain MRI exams ($P=0.11$).

Children who underwent PSP had 4.1 (95% CI 1.0, 16.2) times the odds of completing an interpretable MRI exam compared to children who did not receive PSP ($P=0.0458$) (Table 2). Because age differed significantly between those who did and did not receive PSP, we evaluated the

Table 2 Characteristics of children undergoing unsedated MRIs according to exposure to Preparation and Support Procedure (PSP) intervention

	Successful MRI exam		Unsuccessful MRI exam	
	With PSP	Without PSP	With PSP	Without PSP
Median age (years) (range)	8.9 (5.6-12.9)	10.8 (6.3-12.9)	5.6 (5.4-7.3)	10.9 (6.8-12.9)
No. of successful MRI scans (%)	30 (91%)	27 (71%)*	3 (9%)	11 (29%)
Median number of prior MRIs (range)	1 (0–4)	2 (0–14)	0	1 (0–4)
Median number of prior failed MRIs (range)	0 (0–1)	0 (0–2)	0	0 (0–1)
Median number of prior sedated MRIs (range)	1 (0–3)	2 (0–10)	0	1 (0–3)

*Unadjusted *P*-value=0.0458 for the comparison between successful completion of MRIs with and without PSP, and adjusted *P*-value for same comparison controlling for age *P*=0.0098

association between PSP and completing an interpretable MRI exam using a multiple logistic regression model that controlled for age. After adjusting for age, children receiving PSP had 8.5 (95% CI 1.7, 43.3) times the odds of successfully completing an interpretable MRI exam compared to those who did not receive PSP (*P*=0.0098). Of the 30 children who successfully underwent MRIs with the PSP intervention, 20 (67%) had required sedation/anesthesia for a previous MRI.

Discussion

Procedural sedation can expose children with SCD to dehydration, hypoperfusion, hypothermia, acidosis, hypotension and hypoxia, all of which could trigger a sickle-cell-related adverse event [17–20]. Avoidance of sedation while obtaining adequate diagnostic imaging results is a desirable outcome for young children with SCD. Our results show that the use of preparatory procedures along with supportive measures allowed young children with SCD to undergo MRI testing of the brain and liver, yielding clinically useful results while eliminating the risks associated with the use of sedation. Furthermore, some of these children had undergone prior MRIs with sedation/anesthesia, and with the use of PSP, subsequent MRIs were completed without the use of any type of sedation.

Several alternative techniques to sedation have been employed in infants and young children undergoing painful procedures or imaging studies, such as the use of pacifiers, sucrose, sleep deprivation, guided imagery, play therapy and practice/rehearsal of the imaging study [21–26]. Although practice and support methods during diagnostic imaging studies have been used in different pediatric populations, they have been costly or have been limited by the age group studied and therefore lacked broad application. Most importantly, none of them addressed the SCD population specifically. Hallowell et al. [27] used a full-scale mock MRI machine devoid of magnets to prepare children for the upcoming imaging study; although effective in preparing children for the procedure, the method is too expensive and not a feasible option for most institutions. Khan et al. [28] used equipment such as video goggles during an MRI and moving images projected onto CT scanners to support children during imaging studies. Although these tools, along with preparation by a child life specialist, were effective in decreasing the need for sedation, the technology might not fit within the budget of many hospitals [28]. Pressdee et al. [29] used play therapy methods to prepare children for MR imaging but focused on children 4–8 years old and did not have a control group for comparison. Smart [30] used music and guided imagery audiotape during MRI to provide relaxation to children; while successful in decreasing the need for sedation for the control group, this study also included only children ages 4–8 years, limiting its generalizability. In addition to these limitations, none of these studies was specific to children with SCD—a population that could especially benefit from a procedure that would avoid the need for sedation.

Although use of the PSP method appeared advantageous for our patients, our work has some important limitations. Our retrospective study investigated the utility of a PSP method in young children with SCD who underwent a clinical brain or liver MRI study without the use of sedation/anesthesia. Because a prospective randomized design was not used to evaluate the efficacy of the PSP intervention in reducing the use of sedation/anesthesia, selection bias could have been introduced when referring children for the PSP program. However, gender and prior MRI exposure among children who completed MRI testing successfully were not significantly different compared to those who did not successfully complete an interpretable MRI exam (regardless of PSP participation). In addition, PSP participants did not have a greater number of previous MRI experiences in comparison with non-PSP children. This suggests that gender and previous MRI experiences were not introducing bias into

the selection of participants to the PSP program. The exclusion of children who received upfront sedation/anesthesia without being considered for PSP might also have introduced a selection bias, because these children, owing to their young age or immaturity, may have not had a successful MRI exam without the use of sedation/anesthesia even if receiving PSP. However, because PSP was not even attempted in these 16 children, it is not possible to confirm this hypothesis. Children who participated in this analysis underwent either a brain MRI or liver R2*MRI, lasting approximately 30–60 min, and therefore our findings might not be generalizable to other types of MRI or longer-lasting imaging studies. Our results apply only to children with SCD of similar ages to those of our cohort; therefore these findings should not be generalized to other populations or illnesses. Finally, our study did not prospectively measure process variables of stress reduction and anxiety level, both of which should be included in future prospective study investigating PSP intervention.

Conclusion

Our work to use PSP to allow nonsedated MRI testing in children with SCD is unique. The use of PSP was of significant benefit to children as young as 5 years of age in completing an interpretable MRI exam of the liver or the brain without sedation/anesthesia. The PSP program helped improve coping with MRI diagnostic procedures within the hospital environment while minimizing the risks from sedation known to be associated with children with SCD. The low cost of implementation and ease of use are other important advantages when compared with the other published methods. Prospective randomized studies are warranted to confirm that PSP can successfully facilitate the completion of optimal MRI examinations while avoiding risks associated with sedation/anesthesia in children with SCD.

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