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ORIGINAL ARTICLE

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Family perception of unmet support needs following a diagnosis of congenital coronary anomaly in children: Results of a survey

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Abstract

Background: Long-term outcome data on patients with anomalous aortic origin of coronary arteries (AAOCA) is sparse and they are often managed in a nonuniform manner. There is subjective perception of anxiety and unmet needs in these patients and families.

Methods: An online survey of 13 questions was sent to 74 families of patients with AAOCA between May and October 2015. Descriptive statistics were performed.

Results: A total of 31 (47%) families responded. Of these, 27 expressed the need to interact with other patients/families with AAOCA. The majority were interested in either face-to-face meetings (77%) or online support groups (71%). Regarding content of the meeting, 74% were interested in brief talks by medical personnel/families, 58% suggested informal interactions with families, 55% proposed a structured discussion with a moderator and 39% mentioned fun activities/games. Regarding participants in these meetings, 90% would like to include healthcare providers, 61% suggested including family friends, 58% wished to include psychologists and 16% mentioned including social workers. The families currently use various social media including Facebook (87%), YouTube (39%), Google+ (36%), and LinkedIn (32%). For future online resources, 77% of families would like a Facebook site, an informative website (58%), a blog (52%), or an open forum (29%). The majority of the families (77%) were interested in attending a dedicated AAOCA meeting.

Conclusion: There appears to be an unmet need for family support in those affected by AAOCA, a substantial life changing diagnosis for patients and families. Further research is needed to assess quality of life in this population.

KEYWORDS
anomalous coronary arteries, congenital heart disease, family needs, sudden death, survey

1 | INTRODUCTION

Anomalous aortic origin of a coronary artery (AAOCA) is a congenital anomaly of the origin or course of a coronary artery that arises from the aorta.[1] It is the second leading cause of sudden cardiac death in young young athletes in US.[2] The condition is often asymptomatic and sudden cardiac arrest or death can be its first manifestation.[1] There is little published data regarding long-term management and outcome in patients with these coronary artery anomalies.[3,4] The paucity of data likely relates to several factors but primarily due to the low prevalence of AAOCA in the general population (0.06%–0.9% for anomalous right coronary artery, 0.025%–0.15% for anomalous left coronary artery, and 0.02%–0.67% for anomalous circumflex coronary artery).[1,5–7] This poses a challenge to enroll a large number of patients at any one institution to obtain meaningful research data.

As a result of limited longitudinal data on AAOCA, there is scarce uniformity in physician's approach to the management of this condition.[8]

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Additionally, there has been a recent incidental increase in the rate of diagnosis of these anomalies through various models of cardiac screening, such as a limited cardiac MRI based screening being performed by Angelini et al.[5] in school-aged children. These patients who are diagnosed with coronary anomalies during a screening event are usually asymptomatic and suddenly faced with the diagnosis of a condition that is associated with SCD. Additionally, the natural history has not been clearly demonstrated in patients who have undergone intervention, exercise restriction or no intervention at all.

Parents and caretakers play a vital role in their child's health and are the decision-makers regarding healthcare treatments and procedures. Many parents seek objective data and emotional resources in an attempt to make the best decision for their child.[9–11] The resources utilized often rely on healthcare providers, but also in families or individuals who have experienced a similar situation previously, perhaps even with the same medical condition.[9] The decision-making process in AAOCA is even more difficult given the lack of long-term outcomes and definition of best management strategies. The subjective perception is of a substantial degree of anxiety among families and patients with this disease.

Given limited accessible information paired with nonuniform management strategies, we postulated that these patients and their families have unmet needs following a diagnosis of AAOCA. This survey-based study in a single institution aims to identify resources that these families perceive as potentially being helpful to cope with a diagnosis of AAOCA.

2 | METHODS

All patients evaluated for AAOCA at Texas Children's Hospital were prospectively enrolled following Institutional Review Board approval. An online survey was sent to 74 families to a single parental email address for each family. The survey was sent out twice and the families were instructed to participate only once.

Each survey consisted of 13 questions. Two questions had a single choice response of yes or no and 11 questions were structured in a multiple choice format in which respondents could select multiple answers that applied. The questions addressed various aspects of social support, online tools, and group meetings that families perceived as being useful (Table 1). Descriptive statistics were performed on survey results. Frequency counts and percentages were used for categorical variables.

3 | RESULTS

A total of 31 (47%) families responded to the survey; eight email addresses were invalid. Of those who responded, 27 families expressed that it would be beneficial to meet with other families of patients with congenital coronary anomalies in both face-to-face meetings (77%) and online support groups (71%). The suggested desired frequency for face-to-face meetings to occur was every 6 months (32%), every 3 months (23%), or annually (23%).

There was a variety of topics that families were interested in covering at the proposed meetings: 74% of responding families were interested in brief talks by medical professionals or other patient's families, 58% would like to have informal interactions with other patient's families, 55% were interested in structured discussions with a moderator, and 39% proposed fun activities or games (Figure 1).

In response to the question on who the families would like to see involved in the meetings, 90% responded that they would like the meetings to include physicians and healthcare providers, 61% to include family friends, 58% to include psychologists, and 16% to include social workers (Figure 2).

Several questions addressed potential sources of support in an online format. Responding families stated currently utilizing social media including Facebook (87%), YouTube (39%), Google+ (36%), LinkedIn (32%), and Twitter (10%) (Figure 3). These families would prefer to have the proposed online congenital coronary anomalies resources in the format of a Facebook page (77%), an informative website (58%), a blog (52%), or an open forum (29%) (Figure 4). The patients themselves currently use Facebook (26%), YouTube (45%), Google+ (10%), LinkedIn (3%), and Twitter (22.6%).

Additionally, most families (77%) were interested in attending a scientific meeting dedicated to congenital coronary anomalies and many (68%) would like the meeting to occur annually.

4 | DISCUSSION

Due to the multiple unknowns surrounding AAOCA, patients and families may experience significant anxiety regarding this condition and may thus benefit from dedicated support. The results of this single institution survey suggest that social support in the form of in-person meetings or online websites is a desired resource for families of patients with coronary artery anomalies, specifically AAOCA. This need likely arises from the relatively sparse amount of easily accessible information on AAOCA which is an uncommon disease.[5,8] Parents can often have feelings of guilt and blame with the diagnosis of any congenital heart disease and this needs to be specifically addressed by health care providers.[12] A network of individuals and families with similar diagnoses offers not only resources for emotional support but also of objective and contemporary data that may help in their understanding of this condition.[13,14] These resources can be influential in both coping with this diagnosis and making decisions regarding treatment and exercise activities. Any reliable and informative resource related to the diagnosis and prognosis of a patient's condition can improve the coping ability of the patient and family.[9,13,14]

Furthermore, the creation of networks of patients and families has the potential to effect positive changes in the treatment of these coronary artery anomalies by facilitating communication between patients, families, physicians, and other healthcare providers. This was evidenced in the responses obtained in the survey. These connections may facilitate the gathering of a larger volume of data on patients living with AAOCA, thus providing opportunities to draw reliable conclusions on effectiveness of various treatment plans and medical management decisions. An essential initial effort was the creation of the Congenital Heart Surgeons' Society AAOCA Registry[15] which currently houses retrospective and prospective data on patients with AAOCA from

TABLE 1 Family survey questionnaire

The Coronary Anomalies Program at Texas Children's Hospital is exploring the opportunity to provide our patients and families with a social support structure to facilitate interaction between them and with other support personnel. Your answers to the following survey are important for us to determine the best structure for this support mechanism.	
1. Do you think it would be helpful for your family to interact with other patients and families with coronary anomalies?	• Yes • No
2. What platform for social support would you find useful for you and your family? (mark all that apply)	• Meetings with other families and patients with coronary anomalies • Ability to contact or be contacted by a patient or family on an individual basis • Online support group or forum • Group activities and/or outings • Other (please specify)_____
3. If meetings were to be arranged, how frequently would you want to see them happen?	• Annually • Every 6 months • Every 3 months • Every month • Other (specify) • I'm not interested in face-to-face meetings
4. What would you like to see as part of the meetings? (Mark all that apply)	• Informal interaction between families and patients • Structured discussion with a moderator • Fun activities and games for patients and families • Brief talks by medical personnel and/or families about coronary anomalies • Other (please specify)_____
5. Who do you think should be invited to these meetings besides patients and families with coronary anomalies? (Mark all that apply)	Patients and family friends • Social workers • Psychologists • Physicians and healthcare providers that are part of the Coronary Anomalies Program • Anyone interested in coronary anomalies
6. If meetings were to be arranged, I would likely attend meetings occurring on: (mark all that apply):	• A weekday evening • A weekday morning • Saturday morning • Saturday afternoon
7. If meetings were to be arranged, I would likely attend meetings occurring at: (mark all that apply):	• Main Texas Children's Hospital campus/Texas Medical Center • Texas Children's Hospital West Campus • Different location within the loop • Katy • Memorial City • The Woodlands
8. What online tools and social networks do you regularly use? (Mark all that apply)	• Facebook • Twitter • LinkedIn • Google+ • Flickr • Meetup • Blogger • YouTube • Tumblr • Other (please specify)_____
9. What online tools and networks does your child with the coronary anomaly regularly use? (mark all that apply)	• Facebook • Twitter • LinkedIn • Google+ • Flickr • Meetup • Blogger • YouTube • Tumblr • Other (please specify)_____
10. If an online solution is designed, what structure would you rather see for this?	• A blog with ability to comment • An open forum • A Facebook site • An independent site with information being updated about coronary anomalies • Other (please specify)_____
11. Who should the online support system be open to?	• Patients and families with coronary anomalies being treated at any center in the US • Patients and families with coronary anomalies being treated at Texas Children's Hospital • Physicians and healthcare providers • Everyone interested in coronary anomalies
12. Would you be interested in attending a periodic symposium to update patients and families on advances in coronary anomalies?	• Yes • No
13. If yes, how often would you like to see this happen?	• Every year • Every 2 years • Every 3 years • Other (please specify)

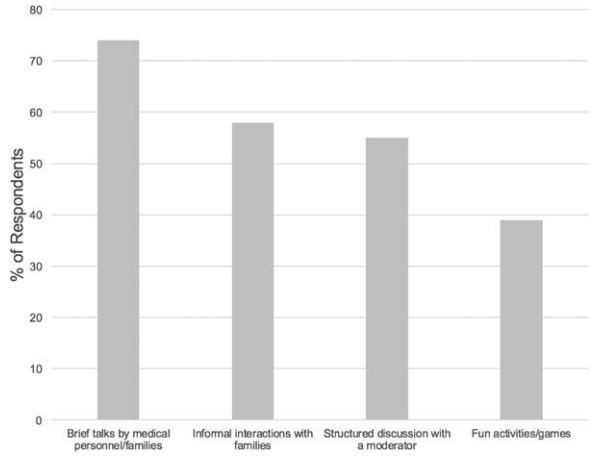

FIGURE 1 What should the content of meetings be

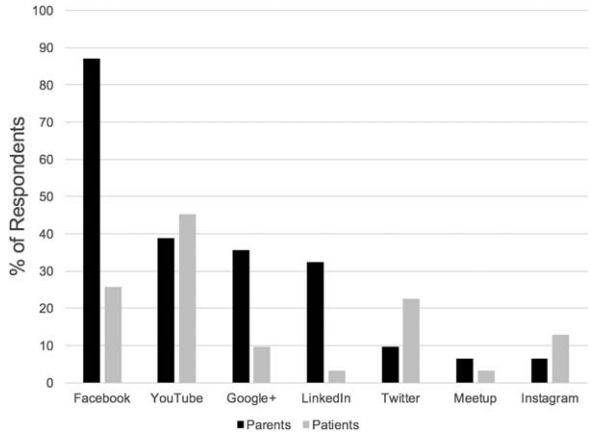

FIGURE 3 Social media being used by the patients and their families

currently 39 North American member institutions. From the patient/family perspective, these efforts can help families understand the implications of this diagnosis and the current available treatment options. Ultimately, this may help parental decision-making process and will lead to improvement in care of patients and families with AAOCA.

Aiming at gathering longitudinal data that would improve our understanding of patients with AAOCA, in December 2012, Texas Children's Hospital created a multidisciplinary Coronary Anomalies Program. The team includes cardiologists, congenital heart surgeons, cardiovascular radiologists, outcomes and research staff, and a clinical algorithm based on the best available evidence was created.[1] Since then, all patients with coronary anomalies have been evaluated and managed in a uniform manner at our institution. Despite heterogeneity among institutions in evaluating and managing patients with AAOCA, data from single institutions and the Congenital Heart Surgeons' Society Registry,[15] will hopefully contribute to meaningful data that will ultimately help these patients and their families. Adopting a uniform approach across the nation will be a paramount step to improve outcomes in this population.

Physical, psychosocial, emotional well-being and occupational adjustment are all crucial parts of a normal healthy life for both patients and families. Alteration in any of the domains can negatively impact quality of life (QOL). Majnemar et al.[16] studied families with children >5 years of age following open heart surgery and found that although the mean QOL scores were normal, parents often reported issues related to attention span, anxiety, learning and development delay in their children. Parental stress level also correlated with child's psychosocial health status.[16] Previous studies using cardiac specific QOL instruments have shown that children and adolescents with congenital heart disease have lower scores compared to healthy controls.[17] Furthermore, lower scores are associated with higher disease severity and increased medical care utilization.[17] However, in a small cohort of patients following AAOCA repair, QOL scores have been reported to be similar to the general population in short and mid-term.[18,19] This likely needs to be investigated further as noncardiac specific questionnaires were used in this study.

Limitations of this study include a small sample size from a single institution that followed a specific algorithm to evaluate and manage these patients. We had a response rate of 47% in this email based survey which is in the average range for survey response rate in questionnaire-based studies,[20] making it generalizable to the families with AAOCA at our institution. This study was performed on a group of families with a variety of AAOCA anatomic variants, hence with varying degrees of risk profile. Depending on the type/severity of the lesions, the patients may have been counseled no intervention, exercise restriction or undergo surgery. Thus, the level of involvement of the families with following recommendations, their accrued knowledge of the condition and unmet needs may be

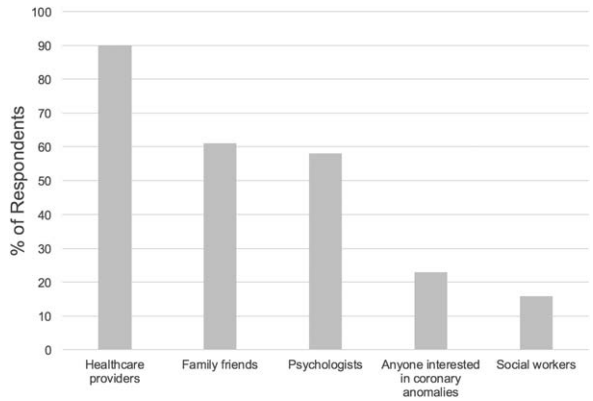

FIGURE 2 Who should be invited to the meetings

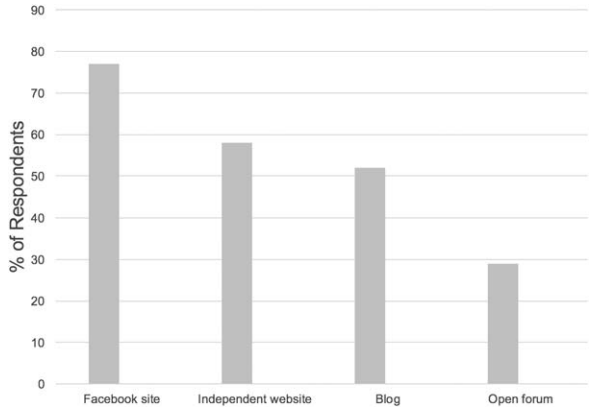

FIGURE 4 Helpful online resources to be developed in the future

heterogeneous within the study population. It has been previously shown that mothers of young patients suffering SCD may have poorer psychological well-being than the fathers.[12] This highlights another limitation of this survey which did not seek to identify which family member answered the survey.

Additional research is needed to identify the opinions and needs of families and patients from a broader range of institutions, as well as to define the impact on perceived QOL of patients and families with and without exercise restriction and following surgery. Additionally, it would be interesting to explore the impact physicians perceive these proposed support groups might have through an additional survey-based study. These can better delineate strategies and support systems that can be successfully implemented in the community. We currently have an ongoing prospective cohort study to assess QOL among patients with AAOCA and their caregivers.

5 | CONCLUSION

Given the lack of long-term outcome data surrounding the management of coronary anomalies, namely AAOCA, there appears to be a defined need for support in these patients and families affected by this diagnosis. Creating reliable online and community based social resources dedicated to these patients and their families may be beneficial, helping them cope with this diagnosis through better understanding and sharing of experiences. Adopting a shared decision-making approach is essential in the care of these patients and their families, and attending to their needs is equally important to be considered by healthcare providers caring for them.

CONFLICT OF INTEREST

The authors have no conflicts of interest to declare.

AUTHOR CONTRIBUTIONS

All the listed authors have made significant contribution in the preparation and review of the manuscript.

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