RESEARCH ARTICLE

# Adult congenital cardiac life-long needs evaluation in a low-middle income country, Pakistan

Laila Akbar Ladak[1], Disty Pearson[2], Kathy Jenkins[3], Muneer Amanullah[4], Waris Ahmad[5], Kaitlin Doherty Schmeck[6], Amy Verstappen[7], Babar Sultan Hasan[8]

## Abstract

**Objective:** Adult congenital heart diseases (ACHD) have distinct health care needs that require life-long care. Limited data is available from low-middle income countries (LMIC). This descriptive study conducted in Pakistan, aimed to assess patients and health care professionals understanding of the needs for ACHD care and the perceived barriers to care.

**Methods:** A telephone survey was conducted of ACHD patients. An e-mail survey was sent to the paediatric and adult cardiologists of five institutions (3 public and 2 private) that provide ACHD services in Pakistan. Descriptive statistics (frequencies, mean ± SD, median) were used for data analysis.

**Results:** A total of 128 ACHD patients were surveyed, 65 (51%) were females with a mean age of 29.4±10.4 years. Atrial septal defect repair was the most common surgical procedure. Mean age at surgery was 25.6±10.49 years, and a surgical follow-up period of 3.8±2.3 years. Majority (n=3, 60%) of the health care professionals (HCPs) responded that 75-100% of the ACHD surgical patients would need lifelong care, yet 10-25% return to their cardiology clinics. Most of the surveyed ACHD patients (89%, n=114) demonstrated a lack of understanding of life-long care after surgery due to not being communicated by their HCPs. Cost and travelling issues were the barriers highlighted by HCPs. Both ACHD patients (96%, n=122) and HCP (100%, n=5) underscored their interest in life-long care.

**Conclusion**: Majority of ACHD patients in Pakistan did not know that life-long follow-up is needed. Education regarding lifelong care for ACHD patients was identified as a means to alleviate the knowledge gap.

**Keywords:** Adult congenital heart disease, Life-long care, Low middle income country, Health care professional

(JPMA 70: 2332; 2020) **DOI: https://doi.org/310.47391/JPMA.013**

## Introduction

Global birth prevalence for congenital heart disease (CHD) is currently estimated at 0.9%, resulting in approximately 1.35 million neonates born with CHD each year worldwide.[1] In high income countries (HIC), advancement in early diagnosis and childhood palliative/reparative surgery has resulted in an 85% rate of survival into adult life. As a result, adults with CHD (ACHD) comprise 22-26% of the total CHD population worldwide.[2] Long term post-operative complications including arrhythmias, heart failure or other obstetric problems affect ACHD patients resulting in multiple hospitalizations.[3] With increasing age, these issues are reported to increase in frequency as well as severity. Life-long care for ACHD and compliance with regular follow-up have been found to be associated with better survival, early identification of morbidities, as well as provision of preventive health care for any potential future issues.[4]

To address the needs of this population, ACHD is an increasingly recognized subspecialty in HIC, with set guidelines and specific training to address the lifelong needs of this population.[5] Furthermore, educating patients and families about their need for life-long care while in paediatric cardiac care, has been defined as a best practice.[2] The most common barrier to continuity of care reported by ACHD patients is their own lack of knowledge about the need for such care.[6] This gap likely reflected in part a historic knowledge gap in the development of congenital cardiology. The extent of the long-term health risks in post-operative CHD did not become fully evident until the 1980's and 1990's, as the first generation of CHD patients to reach adulthood began experiencing unanticipated health problems.[6] This knowledge gap is likely reflected in the large numbers of post-operative adult congenital heart patients, who reported being graduated from paediatric care with no discussion of long term care needs.[7]

A significant burden of CHD exists in low-middle income

---

[1]Charles Perkins Centre, Sydney Nursing School, The University of Sydney, Sydney, Australia; [2]Department of Cardiovascular Medicine, Boston Childrens Hospital, Boston, United States of America; [3]Department of Cardiology, Harvard Medical School, Boston, United States of America; [4]Department of Pediatric Cardiac Surgery, Pediatric Cardiac Services, National Institute of Cardiovascular Diseases, Karachi, Pakistan; [5]Department of Pediatrics Cardiac Surgery, The Aga Khan University, Karachi, Pakistan; [6]Healthcare Sector, PA Consulting Group, Cambridge, Massachusetts, United States; [7]Global Health, University of Barcelona, Barcelona, Spain; [8]Department of Pediatrics and Child Health, The Aga Khan University, Karachi, Pakistan.

**Correspondence:** Laila Akbar Ladak . e-mail: llad8915@uni.sydney.edu.au

countries (LMICs)[8] due to high birth rates. Late presentation and challenges with diagnosis and management further added to patient morbidity.[9] In addition, barriers to proper care included lack of medical infrastructure and knowledge.[5] Pakistan is developing a paediatric cardiac care system in an era in which the need for ACHD care is now well-documented. This offers an opportunity not only to plan for these patients as they develop, also to avoid the "knowledge gap" that resulted in large numbers of ACHD patients in high income countries lost to care.[7,10]

Although barriers to ACHD in high-income countries have been extensively studied,[11,12] little is known from LMIC on barriers to access to appropriate and life-long care. Specifically, no studies have been executed to explore both patient and health care professionals (HCP) knowledge of life-long care needs and their perception to barriers to care provision. This study is the first which aimed to appraise the perception of ACHD patients and HCPs on need for life-long care, barriers to ACHD care/follow-up and their interest in learning about ACHD life-long care and management in a LMIC, Pakistan.

## Methods

This is a survey from five tertiary hospitals in Pakistan providing care to ACHD, including three public sector hospitals.

Approval was sought from the ethics review committee of the study setting. The survey was developed based on the literature search[7,13] and in consultation with the expert professionals in the study team. The survey focused on assessing the perspective of ACHD patients and HCPs on the need for life long care, barriers for regular follow-up, interest in learning about life long care as well as topics of interest and preferred methods (supplementary file 1). Life-long care was defined as the regular annual follow-up under the care of a cardiologist/ACHD following CHD surgery.[5] The patient survey form was translated to Urdu and back translated to English to ensure the translation credibility.

Fifteen HCPs working at the five hospital sites (providing ACHD services) were identified through the Pakistan Paediatric and Adult Cardiology Society websites. The names and contact details of paediatric and adult cardiologists caring for ACHD patients were accessed through Pakistan's Paediatric and Adult Cardiology Society websites.[8] Physicians with high clinical ACHD volume (n=5) from these sites were identified as the site representation, including both paediatric and adult cardiologists. Informed consent and survey questionnaire were e-mailed to these five HCPs followed by a telephone call as applicable.

Post-operative ACHD patients were recruited from one private site, as it has a well-established surveillance system. Patients who were 18 years and above, operated from 2008 till 2016, able to understand English and/or Urdu and without any documented intellectual disability or chromosomal abnormality were identified through the hospital's database system and approached via telephone. A research assistant contacted each patient, provided details about the study, obtained verbal consent and administered the survey. Demographic data was collected from patients during a phone interview while clinical characteristics were retrieved from hospital's medical files.

Data was entered in an SPSS version 22 database. Frequencies, percentages, mean ± SD and/or median (min, max) were calculated for the descriptive analysis.

## Results

### ACHD patients

From 300 eligible patients operated between 2008 and 2016, a stratified convenient sample of 220 (up to 30 patients per year of surgery) were contacted, of whom 150 could be approached and 128 agreed to participate. There was an equal proportion of males and females with a mean age of 29.4±10.3 years (range 18-63 years)(Table). Two-thirds (n=84) of the participants had a high school or University degree, while others were studying. The majority had moderate CHD 123(96.09%), with the most common surgical procedure being repair of an ASD 37 (28.7%), VSD 23(17.8%) or valve replacement without the history of rheumatic heart disease 29(22.65%). The mean age at surgery was 25.6±10.5 years. The mean number of follow-up visits were 1.5 (median=1, range 0-6) during a mean follow-up period (date of surgery till the date of assessment) of 3.8±2.3 years(range 0-8 years).

Majority of respondents 114(89%) thought that they did not need life-long care after ACHD surgery (Figure 1). Most of the participants 126(98%) shared that their physician had discussed post-operative care which entailed healthy life style 59(46%), immediate post-operative care 43(34%, n=43) and medication compliance 26(20%) but not the need for life-long care and regular follow-up.

Participants 122(95%) had the understanding that they should go to see a cardiac surgeon, cardiologist or general physician when it is required or recommended. Patients who had three or more years following surgery 62(47%), were less compliant with follow-up visits.

Upon exploring the reasons for no regular follow-up, participants 112(87%) stated that it was not recommended by their physician or cardiologist while some participants 16(13%) had travel or financial issues (Figure 2).

**Table:** Participants characteristics (n=128).

| **ACHD patients** | **n (%)** |
|---|---|
| **Gender** | |
| Male | 63 (49) |
| Female | 65 (51) |
| **CHD Severity** | |
| Moderate | 123 (96.09) |
| Complex | 5 (3.91) |
| **CHD Surgical procedure** | |
| ASD repair | 37 (28.91) |
| VSD repair | 23 (17.97) |
| AVR/MVR/PVR | 29 (22.66) |
| TOF correction | 29 (22.66) |
| BT Shunt/Glenn Shunt | 5 (3.91) |
| Aortic root replacement/resection of aortic membrane | 2 (1.56) |
| Fontan procedure | 2 (1.56) |
| RV myomectomy | 1 (0.78) |
| **Number of surgeries** | |
| 1 | 111 (87) |
| **Residential place** | |
| Karachi | 114 (89) |
| **Education** | |
| High School | 60 (46.87) |
| University Degree | 24 (18.75) |
| Currently studying High School | 40 (31.25) |
| Currently studying University degree | 4 (3.12) |
| **Occupation** | |
| Business/Service | 34 (27) |
| Home wife | 41 (32) |
| Student | 44 (34) |
| Unemployed | 9 (7) |
| | Mean±SD |
| Age at Assessment (years) | 29.43±10.34 |
| Age at 1st surgery (years) | 25.61±10.49 |
| Follow up years since last surgery (range) | 3.80±2.26 (0-8) |

SD:: Standard Deviation.

Respondents 89(70%) generally preferred a face-to-face follow ups, whereas less preference was given for telephone 37(29%) or e-mail 2(1%) correspondence.

Amongst the survey respondents, 123(96%) were interested to learn about life-long care with a particular focus on awareness about marriage and conception 64(50%), early detection of CHD 51(40%), life style modification 45(35%), medications action/side effects/management 38(30%), leading normal life 45(35%), medications action/side effects/management 38(30%), regular follow-up 32(25%) and post-op care 13(10%). Furthermore, the preferred language of discussion was Urdu and/or English through telephone 52(41%), face to face 45(35%), and written material/flyers/brochures 15(12%).

### Health care professionals:

The participants reported that they were dealing with ACHD patients with atrial septal defect (ASD) 5(100%), ventricular septal defect (VSD) 5(100%), tetralogy of Fallot (TOF) 5(100%), other cyanotic CHDs 5(100%), pulmonary hypertension (PHTN) 5(100%), pregnancy in ACHD patients 5(100%) and rheumatic heart disease (RHD) 4(80%).

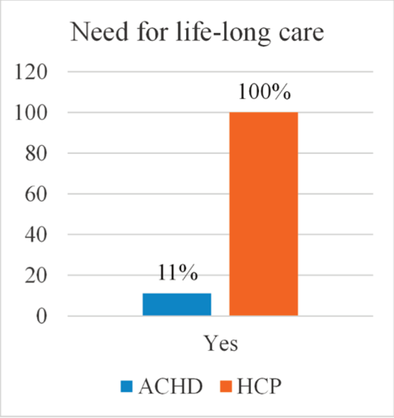

**Figure-1:** Need for life-long care.

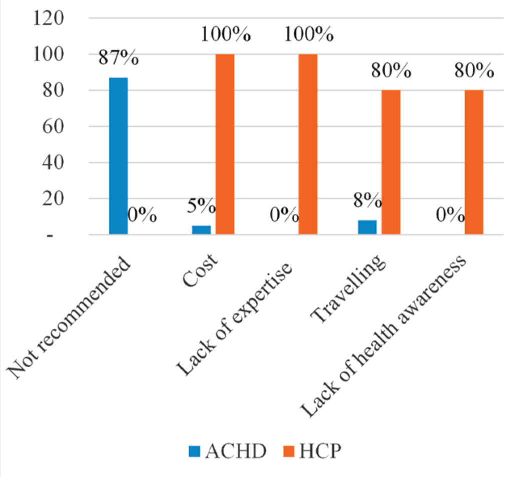

**Figure-2:** Reasons for no regular follow-up.

The majority 4(80%) of HCPs thought that >50% of ACHD patients require lifelong care, while 1(20%) of the HCPs reported that 50-75% of their ACHD patients returned for regular follow-up.

HCPs highlighted major barriers to the provision of care as cost and lack of trained ACHD expertise 5(100%) as well as travelling/transportation and patients' lack of awareness 5(100%) (Figure 2). The majority of HCPs 3(60%) reported that ACHD patients either go to visit paediatricians, pediatric cardiologists, adult general practitioners, or adult cardiologists as there is no established ACHD services and data surveillance system available in Pakistan.

All of the surveyed HCPs were interested in continuing their education related to ACHD. Participants 5(100%) were keen to learn about ACHD care guidelines, while 4(80%) were interested in transition of care, management of pulmonary hypertension (PHTN), Eisenmenger Syndrome, pregnancy and conception. Some 2(40%) were interested to learn about surgical and non-surgical valve replacement and one (20%) was interested in education about heart transplant.

HCPs 5(100%) stated that they prefer to attain the education through webinars, journal articles and in person education, though they 4(80%) also identified written material, partnering with ACHD centers and telemedicine as a preferred medium for continuing education. All the participants 5(100%) suggested written educational material, in person education, peer-to-peer education and support group as potential resources to assist ACHD patients in health literacy in their life-long care and follow-up.

## Discussion

This study identified a critical gap that in a LMIC Pakistan, ACHD surgical patients perceive either no need for life-long care or only when needed after ACHD surgery which was in contrary to the HCPs. The key finding was that the HCPs' identified the reason for no regular follow up was due to resource constraints like travelling, transportation and cost, whereas the ACHD highlighted that they were never told about the need and significance of life long care following their surgery. However, both ACHD and HCPs were interested in learning about life-long care and identified specific topics of educational interest. The study therefore provided insight about potential interventions to improve care of ACHD patients.

Our study findings are in agreement with other studies reporting that regardless of CHD complexity, the common reasons for no regular follow-ups by ACHD patients included "feeling well" and "unawareness of the need for follow-up".[7,10,13] In addition, those who returned for the follow-up did so when referred by another HCP, had new symptoms or wanted to prevent any future problems.[7,10] Patients who had longer gaps in follow-up tended to require urgent cardiac management compared to those who had regular follow-up with a cardiologist or ACHD specialist.[14] Furthermore, ACHD males with less severe CHD complexity, geographical barriers or insurance issues were less compliant in their regular follow ups.[7]

There were discrepancies between the perception of ACHD patients and HCPs regarding the need for life-long care following CHD surgery. While ACHD patients perceived to visit the HCPs when needed, HCPs reported cost of care and transportation as major issues for lack of follow-up. Studies have reported that ACHD surgical patients have poor knowledge about the disease and its care and perceive themselves as "normal" and "cured".[15] This misperception about the life-long care for CHD posed these patients at risk of not complying with the suggested care and not receptive for any co-morbidities or re-operations. Inferring the American Heart Association (AHA) recommendation of annual follow ups even for a mild CHD severity repaired at an adult age,[5] majority of the ACHD study participants had moderate CHD operated at surgery, 25.61±10.49 years and hence should abide with the annual follow ups. Moreover a recent systematic review reported that the long term outcomes of early VSD repair is associated with ventricular and pulmonary function in the adult life.[16]

This study provided the insight that in a low resource country like Pakistan, it is very crucial that the HCPs provide the specific information to these ACHD patients regarding the long term care. Structured educational programmes specific to patient's age, education, cognition, culture, society and beliefs have been reported to increase knowledge about CHD, deteriorating symptoms and improved compliance with follow-up.[17] Lower HRQOL has been reported by female CHD surgical patients in a study from Pakistan.[18] ACHD patients in this study identified gender-specific needs for information, for example, female ACHD patients requested more information regarding marriage and conception. Gender-specific education might trigger interest and thus lead to better compliance with long term care.[19] One study has claimed that there is paucity of data from LMIC in general and Pakistan in particular.[20] Furthermore it has also highlighted that it is very critical to understand the long term needs of the CHD patient population from LMIC's perspective, considering the differences in socio-demographics and the availability of health care resources from HICs.

HCPs in our setting identified education regarding pulmonary hypertension, Eisenmenger Syndrome, pregnancy and transition of care as educational needs necessary to manage late presenting, unrepaired ACHD patients. Such a need might be generalizable to LMICs like Pakistan where late presentation of unrepaired ACHD patients are prevalent. Therefore appropriate education of HCPs, according to the disease spectrum seen in their practice, might lead to more effective results.

ACHD patients highlighted their preferred medium of receiving education through human contact i.e. through verbal communication rather than written. A combination of both have also been reported to be an effective strategy in disseminating health education to patients and their families.[21] Direct face-to-face interaction may be costly and not feasible especially for patients residing in remote parts of the country. Therefore, the use of web or smart phone visual communication applications would be cost effective strategy in improving accessibility and follow-up for such patients. Alternatively, a local nurse or primary health care physician model in collaboration with an ACHD center could be an effective and efficient strategy in coordinating ACHD patients care, early detection of issues and referrals, and education on self-care management.[22]

Late presentation and surgery is common in LMIC due to resource limitations resulting in pre-operative morbidities like severe polycythemia, pulmonary hypertension, infective endocarditis, pneumonia, sepsis, stroke and malnutrition,[23] all of which impact quality of life. The International Society for ACHD (ISACHD) has reinforced the implementation of structured ACHD services to address the long term morbidity issues[2] which has reduced mortality and improve HRQOL in this population. Given the resource constraints and limitation of ACHD services available in Pakistan, capacity building could be enhanced through collaboration between LMIC and HIC. Quality improvement registries with educational webinars, such as the International Quality Improvement Collaborative (IQIC),[24] might be developed to educate HCPs in LMIC regarding ACHD care. Such collaborations would be an important step to improve outcomes for the growing population of ACHD patients in LMIC like Pakistan and address the critical gaps in ACHD care identified in our study.

### Limitations

The study findings must be reviewed in the light of certain limitations. First, the data collection for ACHD patients was from one tertiary care private center that might have attracted participants with a skewed socio-demographic profile and might underestimate the true picture of the needs of ACHD patients in a LMIC, Pakistan. Therefore findings should be generalized to public settings with caution. Secondly, this study used a convenience sample of eligible patients approached through telephone call. Though telephone interviews provided the flexibility to reach the patients residing in far flung areas who otherwise wouldn't have been part of the study, this method of data collection might have restricted participants to discuss sensitive important issues.

## Conclusion

Majority of patients with ACHD were not educated about long-term care. These patients were willing to be educated and followed throughout their lives. Contextual education addressing the needs of the patients and HCPs is much needed.

### Key Messages:

In a LMIC, Pakistan:

- Adult congenital heart disease (ACHD) surgical patients had lack of understanding about life-long care after surgery.
- HCPs highlighted transportation and cost as the barriers to follow up versus patients who indicated that they were never communicated.
- Health education is required to alleviate the knowledge gap for patients.
- Patients as well as health care professionals were keen to learn about life-long care of adult congenital heart disease.

**Disclaimer:** This study has been presented in 12th Global Forum Humanitarian Medicine/3rd Euro-Asian Symposium in cardiology and cardiac surgery, Athens, Greece, 2018 as Oral presentation

**Conflict of Interest:** None

**Sources of Funding:** This project was funded through Department of Paediatrics and Child Health, The Aga Khan University, Pakistan.

## References

1. van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJ, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol 2011; 58: 2241-7.
2. Webb G, Mulder BJ, Aboulhosn J, Daniels CJ, Elizari MA, Hong G, et al. The care of adults with congenital heart disease across the globe: Current assessment and future perspective: A position statement from the International Society for Adult Congenital Heart Disease (ISACHD). Int J Cardiol 2015; 195: 326-33.
3. Verheugt CL, Uiterwaal CS, van der Velde ET, Meijboom FJ, Pieper PG, Sieswerda GT, et al. The emerging burden of hospital admissions of adults with congenital heart disease. Heart 2010; 96: 872-8.
4. Kempny A, Diller GP, Dimopoulos K, Alonso-Gonzalez R, Uebing A, Li W, et al. Determinants of outpatient clinic attendance amongst adults with congenital heart disease and outcome. Int J Cardiol 2016; 203: 245-50.
5. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al. ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease) Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol 2008; 52: e143-e263.
6. Somerville J. 2 - Grown-Up Congenital Heart or Adult Congenital Heart Disease: Historical Perspectives. In: Gatzoulis MA, Webb GD and Daubeney PEF, (eds.). Diagnosis and Management of Adult Congenital Heart Disease. 3rd ed. UK: Elsevier; 2018, p. 10-2.
7. Gurvitz M, Valente AM, Broberg C, Cook S, Stout K, Kay J, et al. Prevalence and predictors of gaps in care among adult congenital heart

disease patients: HEART-ACHD (The Health, Education, and Access Research Trial). J Am Coll Cardiol 2013; 61: 2180-4.

8. Society PPC. Pakistan Pediatric Cardiology Society: For Children born with CHD. 2015.
9. Mohsin SS, Haque A, Shaikh AS, Bano S, Hasan BS. Outcome of infants with unrepaired heart disease admitted to the pediatric intensive care unit: single-center developing country perspective. Congenit Heart Dis 2014; 9: 116-21.
10. Bracher I, Padrutt M, Bonassin F, Santos Lopes B, Gruner C, Stämpfli SF, et al. Burden and impact of congenital syndromes and comorbidities among adults with congenital heart disease. Int J Cardiol 2017; 240: 159-64.
11. Cook SC, Hindes M, Pater C, Maul TM. Measuring hassles in adults with congenital heart disease. Prog Pediatr Cardiol 2016; 43: 99-103.
12. Fernandes SM, Pearson DD, Rzeszut A, Mitchell SJ, Landzberg MJ, Martin GR. Adult Congenital Heart Disease Incidence and Consultation: A Survey of General Adult Cardiologists. J Am Coll Cardiol 2013; 61: 1303-4.
13. Harrison JL, Silversides CK, Oechslin EN, Kovacs AH. Healthcare needs of adults with congenital heart disease: study of the patient perspective. J Cardiovasc Nurs 2011; 26: 497-503.
14. Yeung E, Kay J, Roosevelt GE, Brandon M, Yetman AT. Lapse of care as a predictor for morbidity in adults with congenital heart disease. Int J Cardiol 2008; 125: 62-5.
15. Webb GD. Care of adults with congenital heart disease - A challenge for the new millennium. Thorac Cardiovasc Surg 2001; 49: 30-4.
16. Heiberg J, Redington A, Hjortdal VE. Exercise capacity and cardiac function after surgical closure of ventricular septal defect—is there unrecognized long-term morbidity? Int J Cardiol 2015; 201: 590-4.
17. Goossens E, Fieuws S, Van Deyk K, Luyckx K, Gewillig M, Budts W, et al. Effectiveness of structured education on knowledge and health behaviors in patients with congenital heart disease. J Pediatr 2015; 166: 1370-6. e1.
18. Ladak LA, Hasan BS, Gullick J, Awais K, Abdullah A, Gallagher R. Health-related quality of life in surgical children and adolescents with congenital heart disease compared with their age-matched healthy sibling: a cross-sectional study from a lower middle-income country, Pakistan. Arch Dis Child 2019; 104: 419-25.
19. Al-Khashan HI, Almulla NA, Galil SA, Rabbulnabi AA, Mishriky AM. Gender differences in health education needs and preferences of Saudis attending Riyadh Military Hospital in the Kingdom of Saudi Arabia. J Family Community Med 2012; 19: 172-7.
20. Ladak LA, Hasan BS, Gullick J, Awais K, Abdullah A, Gallagher R. Health-related quality of life in congenital heart disease surgery patients in Pakistan: protocol for a mixed-methods study. BMJ Open 2017; 7: e018046.
21. Marcus C. Strategies for improving the quality of verbal patient and family education: a review of the literature and creation of the EDUCATE model. Health Psychol Behav Med 2014; 2: 482-95.
22. Sillman C, Morin J, Thomet C, Barber D, Mizuno Y, Yang HL, et al. Adult congenital heart disease nurse coordination: Essential skills and role in optimizing team-based care a position statement from the International Society for Adult Congenital Heart Disease (ISACHD). Int J Cardiol 2017; 229: 125-31.
23. Mocumbi AO. The challenges of cardiac surgery for African children. Cardiovasc J Afr 2012; 23: 165-7.
24. Jenkins KJ, Castañeda AR, Cherian K, Couser CA, Dale EK, Gauvreau K, et al. Reducing mortality and infections after congenital heart surgery in the developing world. Pediatrics 2014: 134: e1422-30.