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Factors influencing on SCD patient's family and caregivers in Al-Qunfudah governorate, Saudi Arabia

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ABSTRACT

Background: Sickle cell disease (SCD) is a common hereditary haematological disorder affecting millions worldwide. The current study aimed to improve the quality of life in SCA patients by Periodic psychological assessment and early detection of psychological effects, optimizing the patient's and family's adaptation to the illness. *Methodology*: This study was a survey-based cross-sectional study conducted in Al-Qunfudah governorate, Saudi Arabia. Sickle Cell Disease Burden Interview (SCDBI) vailed questionnaire was used in interviewing the participants. *Results:* A total of 107 participants participated in the study. The vast majority of the samples have SCA alone 50.47%, with one child with SCA 46.73%. *Conclusion:* Early discovery of the psychological effect of SCD is the fundamental goal to achieve to optimize the patient's and family's adaptation to the illness.

Keywords: Sickle cell disease, haematological disorder, caregivers, Al-Qunfudah, Saudi Arabia

1. INTRODUCTION

Sickle cell disease (SCD) represents the most common hereditary hematologic disease. The term sickle cell anemia refers to various diseases, all of which are associated with the presence of abnormal hemoglobin (Hgb) S (Edwards et al., 2005). Heterozygous individuals carry the sickle cell trait, which is generally an asymptomatic hereditary carrier. Homozygous and complex heterozygous individuals have symptomatic disease (Lane et al., 2003). A mutation occurs in the beta-globin gene produces an amino acid substitution from glutamate to valine at position 6 of the beta-globin protein. Hemoglobin S is formed when the altered betaglobin chain (betaS) is incorporated into the hemoglobin tetramer (Canada Hemoglobinopathy Association, 2015). The abnormal hemoglobin polymerizes under conditions like hypoxia and cellular dehydration, leading to deformation of red blood cells. This in turn, leads to

hemolysis and microcirculatory obstruction, tissue ischemia, and necrosis, organ injury, and pain are the sequelae (Pinto et al., 2019). Millions of individuals globally have been affected by SCD, and it is scattered in many parts like Italy, Greece, Central India, and some African countries. Mortality was estimated to rise from 113,000 in 1990 to 176,000 in 2013 (Karimeldin, 2019). In Saudi Arabia, SCD is the commonest inherited disease with prevalence varies significantly in different parts of the kingdom, and it is responsible for a considerable health burden (Bakri et al., 2014). The estimated prevalence for sickle-cell trait ranges from 2% to 27%, and up to 2.6% have SCA in some areas. In the El-Hazmi study, the majority was 267 per 10,000, with the highest prevalence is in the Eastern province, followed by the southwestern regions (Albagshi et al., 2019; Jastaniah, 2011; el-Hazmi et al., 1999).

Al-Qunfudah governorate has the second-highest rates of sickling in KSA (sickle cell trait 12.66% and sickle cell disease 0.3%) according to Alhamdan et al., (2010). A high rate of consanguineous marriage was conducted in Hali, Al-Qunfudhah governorate in the previous study accounts for more than 50%, which makes the population at high risk (Milaat et al., 2018). People with SCD are at great risk for developing multisystem injury associated with significant morbidity and mortality (Edwards et al., 2005; Buchanan et al., 2010; Milaat et al., 2018; Menezes et al., 2013). Throughout life, people with SCD should be considered for neurocognitive evaluation because of Silent cerebral infarct (SCI) present without focal signs and with regular physical examination but with Specific morbidities such as developmental delays, problematic interpersonal relationships, low self-esteem, declining school performance in children, or changes in social role or functions in adults (Yawn et al., 2014; Hijmans et al., 2009; Collins et al., 1998).

SCD is characterized by acute clinical symptoms, such as painful vascular-occlusive episodes, splenic sequestration, acute chest syndrome, neurological, cardiac, renal, bone, growth, and endocrinological complications. That makes the patient seek the emergency services frequently and by chronic complications, affecting many systems (Canadian Haemoglobinopathy Association, 2015; Pass et al., 2000). Previous studies of SCD have reported that the most significant impact on health-related quality of life (HRQL) of patients with SCD and their families is related to the frequency of Painful crises. However, its intensity depends on individual characteristics, family, and the health care provider (Al-Saqladi et al., 2016).

The psychological effect associated with SCD has been thought of mainly because of chronic pain (Morgan et al., 1986). Moreover, psychopathology in patients with SCD has been found to affect the medical outcomes of patients and the frequency of vaso-occlusive crises (Leavell et al., 1983). It is well known that SCD patients have many psychological and social issues like depressive symptoms, school absentees, deterioration in school performance, and enuresis. Besides, anxiety, depression, and psychological complications are established in many studies, and according to Salih KMA, there is a significant effect on HRQL (Karimeldin, 2019; Levenson et al., 2008; Sogutlu et al., 2011).

In a recent study, about half of the SCD children and adolescents patients had either dysthymia (90%) or Major depressive disorder (10%), (Sehlo & Kamfar, 2015). In contrast, some studies failed to find significant levels of depression among SCD patients (Alao & Cooley, 2001; Alhomoud et al., 2018). Moreover, some research suggests that even the families and caregivers are affected psychosocially and might need more social support and psychological care (Adegoke & Kuteyi, 2012; Welkom, 2012). Periodic psychological assessment and early detection of the psychological effect of the disease to optimize the patient's and families adaptation to the illness is vital for the best outcome; on considering the quality of life of sickle cell disease patients, one should never neglect social, emotional, and psychological aspects of the disease (Karimeldin, 2019; National Heart, Lung, and Blood Institute, 2002; Pass et al., 2000).

A significant amount of literature has been published about SCD, but little has been considered quality of life and depression, specifically in Saudi Arabia, and it needs further evaluation (Al Zahrani et al., 2019). Hence the quality of life and psychological aspect of SCD patients has not yet been studied in the Al-Qunfudah governorate. Our study aims to assess the HRQL among SCD patients in the school-age group, summarize the prevalence of depression and anxiety in the sample, and evaluate social support's role.

2. METHODOLOGY

This an observational cross-sectional study was conducted using a self-administered survey distributed through various online social media platforms such as Facebook, Twitter, Instagram, Snapchat, and WhatsApp, focusing on the Al-Qunfudah governorate, Makkah region, Saudi Arabia. Ethical approval was granted from the Umm Al-Qura University ethics committee (ethical number: HAPO-02-K-012-2021-04-661). Sample size calculation for the larger study was determined by power analysis using GPOWER (Graves et al., 2016).

Participants were families and caregivers of children from age of 8–18 years and known to have sickle cell disease (HgbSS) were included. However, children with neurocognitive disorders like cerebral palsy, neurological disorders (previous stroke), or chronic illness such as chronic heart disease, renal disease, metabolic disease, or any other disease were leading to disability and patient

who refused to participate were excluded. The survey was focused on assessing the burden of the illness on caregivers/families: financially and on daily activities based on Sickle Cell Disease Burden Interview (SCDBI), which a valid and reliable tool it was used in various studies to evaluate the disease burden on the families in multiple aspects like the effect of the disease on family activities and the financial burden of the disease (Adegoke & Abioye-Kuteyi, 2012; Ohaeri & Shokunbi, 2002).

The questionnaire was distributed to participants from February 2021 – May 2021 following a stratification process that results in randomly selected groups. The researchers answered any questions about the study that participants had on the spot. Additionally, participants were asked to give their permission and consent for their participation. The data recorded were analyzed using appropriate statistical methods in SPSS v. 23. Frequency was calculated for the categorical variables, and the Chi-square test was used to compare the categorical variables.

3. RESULTS

A total of 107 participants were surveyed among families of children's with SCA and Thalassemia in AL-Qunfuzah, Makkah region, Saudi Arabia. Majority of the children of the families have SCA alone 50.47% compared to both SCA and Thalassemia, which represent 14.02%. Furthermore, SCA carriers represent 35.51% (Figure 1). Majority of the families have one child with SCA 46.73%, while families with two or more represent the lowest percentage (Figure 2).

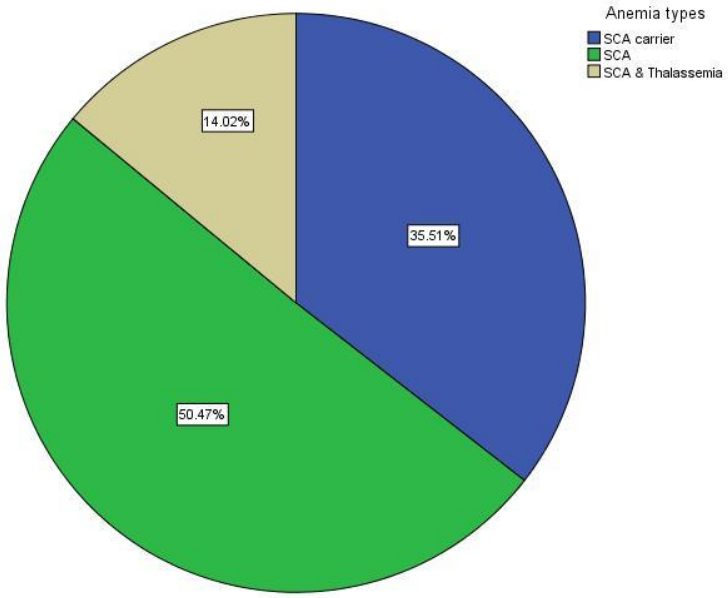

Figure 1 Pie chart of anemia types among children

SCA general impact among caring families was assessed using three scales: never occurred, occurred sometimes, and occurred frequently or always (Table 1). The association between anemia types and SCA general impact among caring families was described in (Table 2). A significant difference was analysed between the influence of the disease financially on the caregivers/families; the effect of the disease on daily activities of the members of the family and Effect of the illness on caregiver/parents' adjustment; coping and their emotions to their child (p-value, 0.253, 0.007, 0.33, respectively) (table 3).

Table 1 Sickle Cell Disease general impact among caring families

Category	Never occurred (%)	Occurred sometimes (%)	Occurred frequently or always (%)
The influence of the Disease financially on the family.	51.7	33.6	14.6
The effect of SCD on daily activities of the members of the family.	34.3	39.0	26.6
Effect of the illness on caregiver/parents' adjustment and coping and their emotions to their child	49.3	29.9	20.7

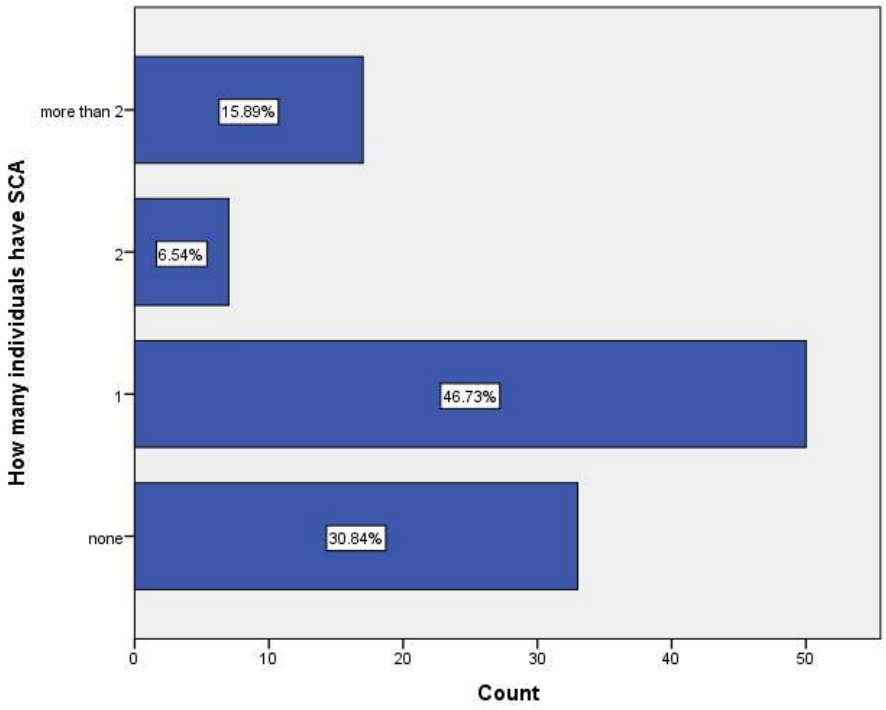

Figure 2 individuals with SCA

Table 2 Sickle Cell Disease general impact among caring families				
Category		Never occurred (%)	Occurred sometimes (%)	Occurred frequently or always (%)
The influence of the Disease financially on the family	Time spend in taking care for the patient lead to Lose income.	42.1	18.7	39.3
	Borrowing money to meet expenditure of the child's illness	66.4	6.5	27.1
	The family basic needs adversely affected by the expenses of diseases	46.7	18.7	34.6
The effect of the disease on daily activities of the members of the family	The time that consumed with the affected patient caused ignorance of other members of the family	28	24.3	47.7
	Disease makes it hard for the patient to help in daily light work of household.	27.1	33.6	39.3
	The disease interfere with home activities	32.7	21.5	45.8
	Difficulty for the parents to engage in activities because of their care for the patient.	49.5	27.1	23.4
Effect of the illness on caregiver/parents' adjustment , coping and their emotions to their child	Trouble in coping with the disease	41.1	15	43.9
	Hard for you to accept the responsibilities for caring of your patient.	56.1	11.2	32.7
	Sadness or unhappy about the child's diseases.	20.6	43	36.4
	Angriness due to child disease.	63.6	18.7	17.8
	Felt stigma because of the disease.	65.4	15.9	18.7

Table 3 the association between anemia types and Sickle Cell Disease general impact among caring families		
Category		p-value
SCD	The influence of the Disease financially on the family.	0.253*
	The effect of the disease on daily activities of the members of the family	0.007*
	Effect of the illness on caregiver/parents' adjustment , coping and their emotions to their child	0.33*

4. DISCUSSION

This study highlights the impact and extent of taking care for children with SCD in their families and caregivers. The caregivers of SCD patients face several challenges financially and on their daily routine family activities and difficulty in coping with their patient (Rattler et al., 2016). In our study, the financial burden of the illness on the families is high; almost 34% of families in this study reported that the family basic needs adversely affected by the expenses of diseases. These findings are consistent with a previous investigational study in a tertiary center in Jeddah, Saudi Arabia, and a study in Nigeria, where the prevalence of SCD is high (Brown et al., 2010; Tunde-Ayinmode, 2007). In addition, approximately 40% of the parents and caregivers reported that Caring for the child interferes with their daily routine family activities. Furthermore, 47% of caregivers reported that the time consumed with the affected patient caused ignorance of other family members.

Concerning coping ability of caregivers and their emotions to the affected child: Caregivers managed differently, as 41% reported never felt difficulty dealing with the child's illness, in contrast, 43% reported as always, moreover one third reported Feeling depressed about the child's condition while, maladjustment and difficulty in coping are known to affect SCD children adversely. Our results indicate that caregivers/parents of children with SCD had significant psychosocial impairment and financial problems. Therefore, social support for caregivers must be considered.

Limitations and recommendations

Our study has possible limitations concerning an insufficient sample size. Moreover, our results are not representative of all Saudi Arabia regions. Therefore, we recommend further investigation.

5. CONCLUSION

The finding of this study demonstrates the significant effect of SCD on caregivers/families of the patients, financially and psychosocially. Moreover, additional attention must be paid to providing convenient services for caregivers and parents of children with SCD.

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Authors Contributions

Both Fawaz Mohammed alzubaidi , Salah Bakry, and Adnan M Alhazmi; were the main participant in writing study introduction, Methodology, discussion and organize the reference part as well as helped in data entry, and the statistical design and analysis. Both Naif Abdullah Almarahabiand Hamzah Zain Sulaiman Alsayed; were participated equally in planed the study as well as helped in data collection. Dr. Mohammed R. Alhayli and Dr. Mohamed A.M. Iesa were helped in the manuscript editing and review. Mohamed A. Elhefny was the General supervisor for the study.

Ethical approval

The study was approved by the Medical Ethics Committee of Umm Al-Qura University (ethical approval code: HAPO-02-K-012-2021-04-661).

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Conflict of Interest

The authors declare that there are no conflicts of interests.

Data and materials availability

All data associated with this study are presented in the paper.

REFERENCES AND NOTES

1. Adegoke SA, Kuteyi EA. Psychosocial burden of sickle cell disease on the family, Nigeria. Afr J Prim Health Care Fam Med 2012; 4(1):1-6.
2. Al Zahrani OS, Mukhtar O, Al Subaie M, Al Howiti WE. Systematic psychiatric assessment of patients with sickle cell disease. Saudi Med J 2019; 40(1):59–65.
3. Alao AO, Cooley E. Depression and sickle cell disease. Harv Rev Psychiatry 2001; 9(4):169-177.
4. Albagshi M, Altaweel H, AlAlwan M, AlHashem H, Albagshi M, AlHabeeb F, AlAmeer H, A Bushehab A· Sickle cell disease awareness among school children in Saudi Arabia. Int J Med Dev Ctries 2019; 28–31.
5. AlHamdan NAR, AlMazrou YY, AlSwaidi FM, Choudhry AJ. Premarital screening for thalassemia and sickle cell disease in Saudi Arabia. Genet Med 2007; 9(6):372–7.
6. Alhomoud M, Gosadi I, Wahbi H. Depression among sickle cell anemia patients in the Eastern Province of Saudi Arabia. Saudi J Med Med Sci. 2018; 6(1):8-12.
7. Al-Saqladi AWM. The impact of sickle cell disease severity on school performance in affected Yemeni children. J Appl Hematol 2016; 7(4):124.
8. Bairwa M, Rajput M, Sachdeva S. Modified kuppuswamy's socioeconomic scale: social researcher should include updated income criteria, 2012. Indian J Comm Med 2013; 38(3):185-6.
9. Bakri M, Ismail E, Elsedfy G, Amr M, Ibrahim A. Behavioral impact of sickle cell disease in young children with repeated hospitalization. Saudi J Anaesth 2014; 8(4):504–9.
10. Brown BJ, Okereke JO, Lagunju IA, Orimadegun AE, Ohaeri JU, Akinyinka OO. Burden of health-care of carers of children with sickle cell disease in Nigeria. Health Soc care community. Wiley Online Library. 2010; 18(3):289–95.
11. Buchanan G, Vichinsky E, Krishnamurti L, Shenoy S. Severe Sickle Cell Disease—Pathophysiology and Therapy. Biol Blood Marrow Transplant 2010; 16(1):S64-S7.
12. Canadian Haemoglobinopathy Association. An introduction to sickle cell disease. In: Consensus Statement on the Care of Patients with Sickle Cell Disease in Canada Version 2 0 Ottawa; 2015 pp 7
13. Collins M, Kaslow N, Doepke K, Eckman J, Johnson M. Psychosocial interventions for children and adolescents with sickle cell disease (scd). J Black Psychol 1998; 24(4):432–54.
14. Edwards CL1, Scales MT, Loughlin C, Bennett GG, Harris-Peterson S, De Castro LM, Whitworth E, Abrams M, Feliu M, Johnson S, Wood M, Harrison O KA. A brief review of the pathophysiology, associated pain, and psychosocial issues in sickle cell disease. Int J Behav Med 2005; 12(3):171–9.
15. el-Hazmi MA ,Warsy AS. Appraisal of sickle-cell and thalassaemia genes in Saudi Arabia. East Mediterr Heal J 1999; 5(6):1147-53.
16. Graves JK, Hodge C, Jacob E. Depression, Anxiety, and Quality of Life in Children and Adolescents with Sickle Cell Disease. Pediatr Nurs 2016; 42(3):113-9, 144.
17. Hasan SP, Hashmi S, Alhassen M, Lawson W, Castro O. Depression in sickle cell disease. J Natl Med Assoc 2003; 95(7):533–537.
18. Hijmans CT, Grootenhuis MA, Oosterlaan J, Last BF, Heijboer H, Peters M, Fijnvandraat K . Behavioral and emotional problems in children with sickle cell disease and healthy siblings: Multiple informants, multiple measures. Pediatr Blood Canc 2009; 53(7):1277–83.
19. Jastaniah W. Epidemiology of sickle cell disease in Saudi Arabia. Ann Saudi Med 2011; 31(3):289–293
20. Karimeldin MA Salih. The impact of sickle cell anemia on the quality of life of sicklers at school age. J Fam Med Prim Care 2019; 8(2):468–471.
21. Klaufus L, Verlinden E, van der Wal M, Kösters M, Cuijpers P, Chinapaw M. Psychometric evaluation of two short versions of the Revised Child Anxiety and Depression Scale. BMC Psychiatry 2020; 20(1):47.
22. Lane PA, Buchanan GR, Desposito F, Pegelow CH, Vichinsky EP, Woods Gerald M. Health supervision for children with sickle cell disease. Pediatrics 2003; 111(3):710–1.
23. Leavell S, Ford CV. Psychopathology in patients with sickle cell disease. Psychosomatics 1983; 24(1):23–5
24. Levenson JL, McClish DK, Dahman BA, Bovbjerg VE, de A Citero V, Penberthy LT, Aisiku IP, Roberts JD, Roseff SD,

Smith WR. Depression and anxiety in adults with sickle cell disease: the PiSCES project. Psychosom Med 2008; 70(2):192-6.

25. Madani BM, Al Raddadi R, Al Jaouni S, Omer M, Al Awa MI. Quality of life among caregivers of sickle cell disease patients: a cross sectional study. Health Qual Life Outcomes 2018; 16(1):176.
26. Menezes AS, Len CA, Hilário MO, Terreri MT, Braga JA. Quality of life in patients with sickle cell disease. Rev Paul Pediatr 2013; 31(1):24-9.
27. Milaat WA, Ibrahim NK, Albar HM. Reproductive health profile and circumcision of females in the Hali semi-urban region, Saudi Arabia: A community-based cross sectional survey. Ann Saudi Med 2018; 38(2):81–9.
28. Morgan SA, Jackson J. Psychological and social concomitants of sickle cell anemia in adolescents. J Pediatr Psychol 1986; 11(3):429-40
29. National Heart, Lung, and Blood Institute. *Management and Therapy of Sickle Cell Disease*. 4th ed. Bethesda, MD: National Institutes of Health; 2002. In press
30. Ohaeri JU, Shokunbi WA. Psychosocial burden of sickle cell disease on caregivers in a Nigerian setting. J Natl Med Assoc 2002; 94(12):1058-70.
31. Pass KA, Lane PA, Fernhoff PM, Hinton CF, Panny SR, Parks JS, Pelias MZ, Rhead WJ, Ross SI, Wethers DL, EL 2nd. US newborn screening system guidelines II: follow-up of children, diagnosis, management, and evaluation. Statement of the Council of Regional Networks for Genetic Services (CORN). J Pediatr 2000; 137(4):S1-46.
32. Pinto VM, Balocco M, Quintino S, Forni GL. Sickle cell disease: a review for the internist. Intern Emerg Med 2019; 14(7):1051-1064.
33. Rattler TL, Walder AM, Feng H, Raphael JL. Care coordination for children with sickle cell disease: A Longitudinal Study of Parent Perspectives and Acute Care Utilization. Am J Prev Med Elsevier 2016; 51(1):S55–61
34. Sehlo MG, Kamfar HZ. Depression and quality of life in children with sickle cell disease: The effect of social support. BMC Psychiatry 2015; 15(1):1–8.
35. Sogutlu A, Levenson JL, McClish DK, Rosef SD, Smith WR. Somatic symptom burden in adults with sickle cell disease predicts pain, depression, anxiety, health care utilization, and quality of life: the PiSCES project. Psychosomatics 2011; 52(3):271–9.
36. Tunde-Ayinmode MF. Psychosocial impact of sickle cell disease on mothers of affected children seen at University of Ilorin Teaching Hospital, Ilorin, Nigeria. East Afr Med J 2007; 84(9):410–419.
37. Varni JW, Seid M, Kurtin PS. PedsQL 4.0: reliability and validity of the Pediatric Quality of Life Inventory version 4.0 generic core scales in healthy and patient populations. Med Care 2001; 39(8):800-12.
38. Welkom JS. The impact of Sickle Cell Disease on the family: an examination of the illness intrusiveness framework 2012.
39. Yawn BP, Buchanan GR, Afenyi-Annan AN, Ballas SK, Hassell KL, James AH, Jordan L. Lanzkron SM, Lottenberg R, Savage WJ, Tanabe PJ, Ware RE, Murad MH, Goldsmith JC, Ortiz E, Fulwood R, Horton A, John-Sowah J. Management of Sickle Cell Disease: Summary of the 2014 Evidence-Based Report by Expert Panel Members. AMA. 2014; 312(10):1033–48.