EVIDENCE BRIEF

Newborn screening
(DCP4 ID: MNH08)

FairChoices
DCP Analytic Tool

Sickle cell screening and management

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Date updated: 2.12. 2021

Description of condition and intervention

Sickle cell disease is a hereditary disorder of hemoglobin (Hb), that occurs due to inheritance of two abnormal Hb genes, which leads to the production of HbS. Sickle cell anemia develops when the individual has homozygote (Hb SS) clinical phenotype. Chronic hemolytic anemia and recurrent vaso-occlusion are common manifestations of this condition. Occurrence of infections like malaria and pneumococcal in individuals with sickle cell disease increases the likelihood of morbidity and mortality in those affected. This is a serious public health challenge in Sub-Saharan Africa which had majority of newborns with sickle cell disease as compared to other parts of the globe (Kuznik et al 2016).

In this evidence brief, we assess the effect and cost of sickle cell screening and management intervention that is being analysed in FairChoices: DCP Analytical tool.

Sickle cell screening and management

Intervention attributes

Type of interventions

Chronic management care

Delivery platform

First-level hospital

Equity

In addition to considerations like cost-effectiveness and health systems factors, dimensions of equity can be relevant for priority setting. The opportunity for a long and healthy life varies according to the severity of a health condition that individuals might have, so there are inequities in individuals' opportunities for long and healthy lives based on the health conditions they face. Metrics used to estimate the severity of illness at an individual level can be used to help prioritize those with less opportunity for lifetime health. FairChoices: DCP Analytics Tool uses Health adjusted age of death (HAAD), which is a metric that estimates the number of years lived from birth to death, discounting years lived with disability. A high HAAD thus represents a disease less severe in terms of lifetime health loss, while a low HAAD represents a disease that is severe on average, causing early death or a long period of severe disability. It is also possible to estimate the distribution of HAAD across individuals with a health condition. FairChoices shows for each intervention an average HAAD value of the conditions that are affected by respective interventions that have health effects. Additionally, a plot shows HAAD values for around 290 conditions (Johansson KA et al 2020).

Time dependence

Moderate level of urgency. Treatment outcomes not highly affected by some days of delay.

Population in need of interventions

Table 1: Population in need of interventions

Intervention	**Treated population**		**Affected population**		**Disease state addressed**
	Treated age	Treated fraction	Affected age	Affected fraction	
Sickle cell screening and management: Newborn screening	births; both genders	births	No effects for diagnostic intevention		Sickle cell disorders
Sickle cell screening and management: Prophylaxis against bacterial	births; both genders, incidence	1	0 to 99years	1	Sickle cell disorders

infections and malaria					

Intervention effect and safety

Table 2: Effect and safety of interventions for sickle cell management

Effect of intervention		Certainty of evidence
Mortality Sickle cell screening and management: Prophylaxis against bacterial infections and malaria	0.98 relative reduction in neonatal mortality (assumed)	Low See appendix

Model assumptions

Table 3: Summary of model parameters and values used in FairChoices – DCP Analytical Tool

Category	Model parameter	Notes
Interventions	Sickle cell screening and management	
Cost calculation		
Treated population	See Table 1	Global Burden of Disease Study 2019
Gender		
Age		
Treated fraction		
Effect calculation		
Affected population	Those with condition	
Affected gender	See Table 1	
Affected fraction age		
Affected fraction		
Comparison	No intervention	
Mortality Reduction (RRR)	0.98 (assumed)	

Intervention cost

The total cost for sickle cell disease screening was 15.36 PPP per infant in a specified population in 2011 in Angola. The total treatment cost for prophylaxis against bacterial infections and malaria is estimated to be 966.067 PPP in 2011 in Angola. The costs were converted to local currency, 992.34 AOA and 62413.54 AOA, respectively, using world bank exchange rates (McGann PT et al 2015).

Commented [SA1]: A Cost-Effectiveness Analysis of a Pilot Neonatal Screening Program for Sickle Cell Anemia in the Republic of Angola (nih.gov)

References

Kuznik et al 2016: Kuznik A, Habib AG, Munube D, Lamorde M. Newborn screening and prophylactic interventions for sickle cell disease in 47 countries in sub-Saharan Africa: a cost-effectiveness analysis. BMC Health Serv Res. 2016 Jul 26;16:304. doi: 10.1186/s12913-016-1572-6. PMID: 27461265; PMCID: PMC4962462.

Johansson KA et al 2020: Johansson KA, Coates MM, Økland JM, Tsuchiya A, Bukhman G, Norheim OF, Haaland Ø. Health by disease categories. Distributional Cost-Effectiveness Analysis: Quantifying Health Equity Impacts and Trade-Offs. 2020 Sep 30:105.

McGann PT et al 2015: McGann PT, Grosse SD, Santos B, de Oliveira V, Bernardino L, Kassebaum NJ, Ware RE, Airewele GE. A Cost-Effectiveness Analysis of a Pilot Neonatal Screening Program for Sickle Cell Anemia in the Republic of Angola. J Pediatr. 2015 Dec;167(6):1314-9. doi: 10.1016/j.jpeds.2015.08.068. Epub 2015 Oct 23. PMID: 26477868; PMCID: PMC4662897.

Appendix

Literature Review for effectiveness & safety

This literature search is an example of Level 1 search for intervention inputs taken from DCP3 or generated in an ad hoc manner (e.g., quick google search found one study of cervical cancer screening cost-effectiveness that was used to create an effectiveness parameter for that intervention).

Level of evidence of efficacy studies:

1. low (expert opinions, case series, reports, low-quality case control studies)
2. moderate (high quality case control studies, low quality cohort studies)
3. high (high quality cohort studies, individual RCTs)

4. very high (multiple RCTs, meta-analysis, systematic review, clinical practice guidelines)