

Yesterday, Today and Tomorrow in the World of Sickle Cell

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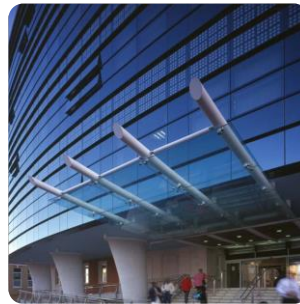
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Background

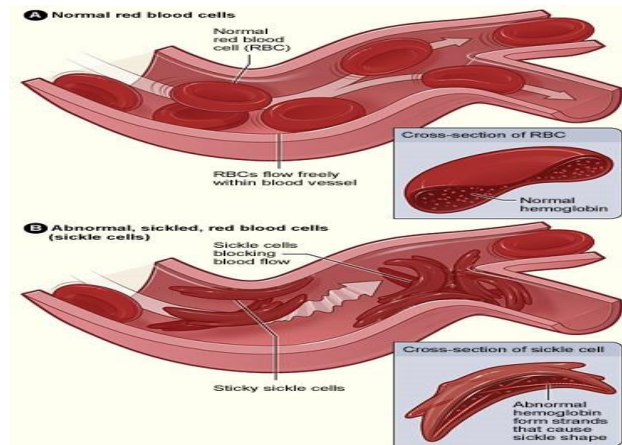
- Sickle Cell Disease is an inherited condition affecting the red blood cells (RBC's).
- A result of mutation of the 6th codon of the gene for beta globin. (Valine substituted for Glutamic acid).
- The first disease to be described at a molecular level (in 1949 Pauling).
- Platt et,al (1994) stipulated that 'Information on life expectancy and risk factors for early death among patients with sickle cell disease (sickle cell anaemia, sickle cell-haemoglobin C disease, and the sickle cell-beta-thalassaemia) is needed to counsel patients, target therapy, and design clinical trials'.
- Reluctance of paediatric providers and young adults with SCD to transfer to adult primary care and specialty care has been based on several factors, including the perception, with some factual basis, that the transfer may result in an increase in morbidity and mortality. (Brousseau 2010 in De Baun et,al 2012)

Background

What Is Sickle Cell Disease?

NHLBI, NIH

National Heart, Lung, and Blood Institute - NIH



Aim of Study

- Semi-structured patient interviews exploring the experience of care, and transition from children's to adult's services for sickle cell patients at Kings College Hospitals Foundation Trust with NHS ELECT

(August 2016– June 2017)

King's models for Transition of care

core elements: (APIE) Assess Plan Implement and Evaluate

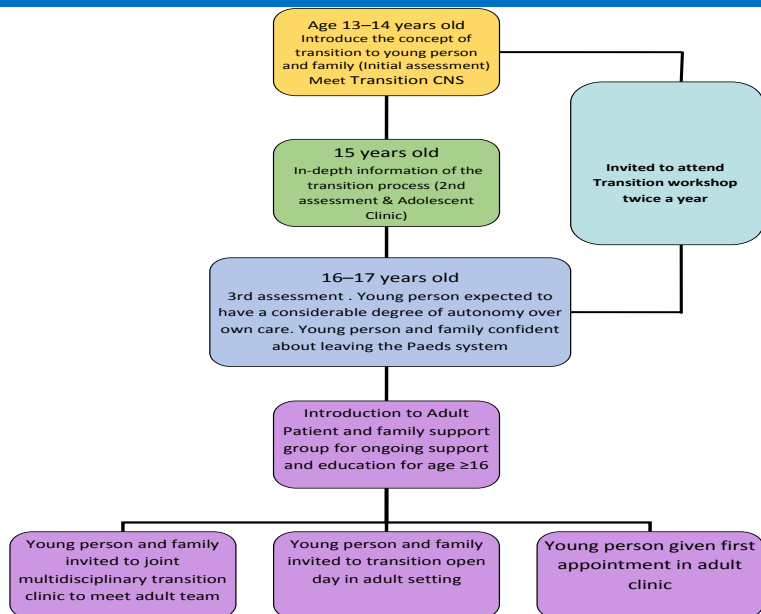
- 1. Transition Policy
- 2. Transition tracking and monitoring
- 3. Transition readiness
- 4. Transition planning
- 5. Transfer of care
- 6. Transfer completion

In Collaboration with Kings the NHS Elect aimed to provide qualitative information from patients to support the development of these core elements.

NHS ELECT 2017, Dr Chakravorty and CNS Giselle Padmore-Payne

Core elements :Adapted from both the American Academy of Pediatrics (<http://www.AAP.org>) and the National Center for Medical Home Implementation (<http://www.gottransition.org>) (2011)

Sickle Cell Transition Pathway



Objectives

- The objective of this audit is compare King's current practice against NICE Clinical Guideline CG50 .Acutely ill patients in hospital and identify and implement actions required to comply with the guideline.

King's team's proposal/brief clearly set out that its commitment

- 'For young people (YP) reaching adolescence, a seamless transition of care from paediatric to adult services is paramount.
- Move to adult services is often fraught with anxieties in YP and carers, as they find it harder to part with familiar paediatric teams.

(Chakravorty 2016)

(NHS ELECT 2016)

Sample and data collection method

Sample

- Sample size: **10 patients in total**
- Number of cases audited: **10**
- Inclusion criteria: **16-24 , HbSS, HbSC, already using London Ambulance Service, Already had an admission to Adult Accident and Emergency Department and Admissions to Adult wards .**
- How was the sample identified: **Through the Paediatric/Adolescent clinic, Adult clinic and Social Media advertisement within the trust**

Data Collection Method:

- Data collection type: **Initially a mixed methods approach was proposed that comprised of the following components:**
- **Semi-structured Questions via telephone**

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Sample and data collection method

- **pathway mapping** session with users and carers; a staff session; then a joint workshop of users, carers and staff to work through the pathway and agree improvements/recommendations together
- **themed discussions** with young people at existing events or meetings or through a bespoke gathering
- **1:1 conversations** with patients in either outpatient or inpatient settings
- **engagement through social media** to promote engagement sessions
- Recruitment Challenges : difficulty with patient attendance, therefore, thinking outside the box approach and study had to be extended

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Key Findings:

- **Pathway mapping** session with users and carers was well publicized, and scheduled to be held on the Denmark Hill site, however only 1 patient attended.
- A series of other **drop in sessions** were scheduled during clinics to try and capture more, again only a very small number of patients attended, and not all had very much that they wanted to share. There were no existing user forums scheduled.
- This evidenced the need for a **flexible approach**, and an appreciation that this patient group did not wish to attend unnecessary meetings on the hospital site. The period of the project was extended, with an initial focus on patient feedback, ahead of staff sessions.
- **1:1 semi-structured interviews** were found to be the best method, particularly by phone.

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Results

- In total **10 patients** talked to us about their experiences. All <24 years of age. There was a mixture of patients who had transitioned, were about to transition, or who were still under the paediatricians without a planned date.
- Categories '**what worked**', '**worked well**' and '**worked less well**' headings of the **ambulance; the ED; paediatric inpatients; adult inpatients and transition.**
- Each patient was asked if they had any **suggestions** about how services could be improved.

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Individual reflection

- **Scared of transition. Doesn't trust the nurse's** description and reassurance of what it will be like, as her current experience of the adult service is poor.
- Currently **anxious** about what will happen on the adult wards. **Liaison with paed**s would be really helpful so that both on the same page. Needs to be a basic understanding about what is going on. If I am staying for a long time I would want to be on paediatrics than in haematology adults. The A&E is massive. Horrible environment – the environment is horrible
- We want **some sort of separation**. E.g. my last stay was ITU/one day stay but there was a drug addict to calm down. Then another elderly lady. Someone was having a seizure – not ideal
- Would **want to feel reassured** that the adult service was geared towards young adults
- **Suitable environment** for young adults, wifi, social media, TVs etc. (similar to other units – TYA cancer floor at UCLH)
- **One place for admissions** to know where I would be going, and not to be moved around too much. It would be good if I only need to give my details once, and for **staff to know me.**
- NB despite asking several times about how to make transition easier, the key thing that came up many times was about the experience of being on adult wards, rather than transition itself (see next slide).
- 'Giselle has been really good at helping me transition. She has been really great at managing me pro-actively, and giving me information. If I didn't have Giselle it wouldn't have gone so well'. We need a nurse to help with the process.

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Key Themes Overall

- **Experiences** are better when the YP is **accompanied** by a family member, and not reliant on hospital transport.
- **advocacy** came out strongly – its presence or absence had a real impact on experience.
- **Positivity about their experience of the paediatric service**, on of the haematologists when under their care.
- All YP talked about the **varying levels of knowledge, understanding, and values and behaviours of staff outside of paediatrics/haematologists**. Several referenced that education would be helpful.
- Patients **felt they weren't listened to by health care professionals outside of the paediatric team** regarding who's care they are currently under, or their condition and management.
- YP **upset and frustrated** that their notes and **(standard or tailored) care plans weren't available and that they had to repeat information several times**. Several said that when they tried to explain their treatment plan they weren't listened to and weren't seen as 'expert patients and carers'.
- **Pain management** is clearly a complex issue, linked to the themes above, and will probably require further exploration. NHS ELECT 2017

Recommendations

A small debrief meeting was facilitated via telephone conference in order to agree way forward, dissemination and discussion with the wider team/staff

Organisation of Stakeholders meeting in upcoming Months (September)

Consider and discuss the individual responses and overall themes of the report, and agree core recommendations and Strategic Approaches

Agree how to feedback to service users/participants and families

Publish and share on social media eg STSTN, Sickle Society and National Media

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Dr Chakravorty

CNS Giselle Padmore-Payne

Summary

Poor transition of care in SCD → Associated with increased mortality.

Brousseau et al provided an indirect measure of increased morbidity in the age group most affected by transfer of care, 18- to 30-year-olds.

The concerns that have been expressed by Young People in this mixed method study reflects similar if not the same concerns. Prior to this study transition in previous studies were well-founded and experienced in adulthood by a number of participants.

The ability of youths with SCD to participate fully in society and live productive lives as adults depends on their ability to navigate the transition period successfully. (Treadwell et,al 2015)

Many patients felt and continue to feel that they are poorly treated and historically they found that the adult providers knew little about sickle cell. Today however, there are more adult providers specialising in sickle cell/haemoglobinopathies.

Tomorrow

No single scientific advancement has contributed to the increase in survival; however, several noticeable improvements greatly increased the survival rate of infants and children with SCD. (Quinn et,al 2004)

Due to advancements in today's treatments and medications .The life span for children with SCD has increased dramatically in the past 50 years.(Hasell et,al 2010)

The Care Act 2014 places a "duty on local authorities to conduct transition assessments for children, children's carers and young carers where there is a likely need for care and support after the child in question turns 18 and a transition assessment would be of 'significant benefit'".

Our aim for tomorrow is to enable , empower, enlighten, and facilitate transition through education and systematic approaches collectively for service users, families and health care professionals, we will help to overcome the aforementioned barriers and reduce the mortality and morbidity rates for young adults who live with Sickle Cell Diseases.

Standards for Practice

NHS Elect was commissioned to work with Kings College Hospital in August 2016 to gain insights from Young People with SCD into their current experience of their care, with particular focus on transition to adult services

“Ensuring a seamless transfer is one of the greatest challenges facing both children’s and adult services” RCN 2004.

UK national standards for sickle cell disease in childhood: using audit to improve clinical care (Hann et al 2012)

Lost in Transition ‘Moving young people between child and adult health services’ Royal College of Nursing (RCN 2013)

Transition from children’s to adults’ services for young people using health or social care services (NICE guideline 2016) nice.org.uk/guidance

The Children and Families Act 2014 and the Care Act 2014

References

- Brousseau DC, Owens PL, Mosso AL, Panepinto JA, Steiner CA. 2010 Acute care utilization and rehospitalizations for sickle cell disease. JAMA. 2010;303(13):1288–1294pmid:20371788
- Children and Families Act 2014 Transition from children’s to adult services – early and comprehensive identification <http://www.scie.org.uk/care-act-2014/>
- DeBaun, M.,and Telfair, J.,Transition and Sickle Cell Disease Paediatrics 2012;130;926; originally published online October 1, 2012;
- Hassell KL. Population estimates of sickle cell disease in the U.S. Am J Prev Med. 2010;38(suppl 4):S512–S521pmid:20331952

References

- Platt OS, Brambilla DJ, Rosse WF, Milner PF, Castro O, Steinberg MH, Klug PP. Mortality in sickle cell disease. Life expectancy and risk factors for early death. N Engl J Med. 1994 Jun 9; 330(23):1639-44.
- Quinn CT, Rogers ZR, Buchanan GR. Survival of children with sickle cell disease. Blood. 2004;103(11):4023–4027pmid:14764527
- Royal College of Nursing RCN (2013) Lost in Transition 'Moving young people between child and adult health services' Royal College of Nursing publications.
- Reiss J, Gibson R. Health care transition: destinations unknown. Paediatrics. 2002;110(6 pt 2):1307–1314pmid:12456950

References

- Royal College of Nursing (2004). "Ensuring a seamless transfer is one of the greatest challenges facing both children's and adult services". RCN publications.
- Transition from children's to adults' services for young people using health or Social care services (NICE guideline 2016) nice.org.uk/guidance
- Treadwell,M., Johnson,S.,Sisler,I., Bitsko,M., Gildengorin,G.,Medina,R., Barreda,F., Major,K., Telfair,J., and Smith,W.R.,Development of a sickle cell disease readiness for transition assessment. Int J Adolesc Med Health 2015
- UK national standards for sickle cell disease in childhood: using audit to improve clinical care (Hann et al 2012)

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NHS Elect Lisa Godfrey and Team

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Dr Chakravorty Transition Lead and Haemoglobinopathies Consultant

Dr Sarah Bennett Adult Haemoglobinopathies Consultant

Dr Kate Gardner Team Research Fellow for Haemoglobinopathies

Marlene Allman Senior CNS for Adult Haemoglobinopathies

Last but not least Our USA counterparts and Haemoglobinopathies Team

Without everyone's input and assistance service improvement cannot be facilitated.