



Using a Nursing Model in the management of a patient with McCune Albright Syndrome

Caroline Jagger, Manchester Royal Infirmary, March 2013

Background of the Disease

- McCune Albright syndrome and Fibrous Dysplasia are a rare genetic skeletal disorder in which normal bone structures and marrow are replaced by a benign fibrous tissue. It is a non-inheritable genetic disease that can be limited to one or many bones, it is more prevalent in females. It can cause pathological fractures and severe bone pain for which bisphosphonates are an effective treatment (Muriel et al 2003, Chapurlat 2008, Dumitrescu 2008).

Past medical history of the patient

- Precocious Puberty, Café au lait skin pigmentation
- Multiple pathological fractures in childhood requiring surgical management
- Cholecystectomy 1996
- Thyrotoxicosis 1997
- Mastectomy for recurrent Fibrocystic disease, Mastectomy for ductal carcinoma 2011
- Carpal Tunnel release 2006
- Hysterectomy, bilateral oophorectomy 2010

Presenting problem

- Patient 48 year old female attends the outpatient clinic every 6 months
- Recently increased pain in skull preventing sleep and causing severe pain,
- Plan: bloods bone profile, Urea and Electrolytes, Parathyroid Hormone, Vitamin D
- Thyroid Function discussion with Consultant regarding pain, to have CT scan skull, infusion of Zoledronic acid if blood results within the normal range.

Specialist Nurse role

- Explain to patient the length of time blood results will take and act on those results
- Give contact number of helpline
- Ask the patients availability to attend the Hospital
- Liaise with Patient and Programmed investigation unit to arrange a mutually convenient time for treatment Prescribe Zoledronic acid/ Colecalciferol

Multi-disciplinary team

- In order to arrange the admission there are many different departments to liaise within the hospital
- Consultant
- Pharmacy
- Phlebotomists
- Biochemistry
- Secretaries
- Programmed Investigation Unit Administrator

Nursing Model

- As the patient's needs and requirements are at the forefront of care, it is essential that the holistic needs are met for the patient. There are several frameworks that can be utilised to ensure these needs are met. The Roper, Logan and Tierney model (1980) is one that is synonymous with nursing practice.

Roper, Logan and Tierney (1980) Nursing model

- Designed as a framework for nursing care and practice, in which it ensures the holistic care of the patient by encompassing 12 activities of daily living, Maintaining a Safe Environment, Communicating, Breathing, Eating and Drinking, Eliminating, Personal Cleansing and Washing, Controlling Body Temperature, Mobilising, Working and Playing, Expressing Sexuality, Sleeping and Dying. The model also encompasses the Physical, Psychological, Sociocultural, Environmental and Economic aspects of the patient.

Maintaining a safe environment

- When attending outpatients, move chairs in consulting room to promote equity and objects being a hazard.
- On the Programmed investigation Unit ensure drip stands are safely stored, as the patient has motorised wheelchair.
- To promote independence as much as possible, advise the patient to wear appropriate clothing, making it easy to insert a cannula for the infusion. Using ANTT reduces the risks of infection to the patient.

Communication

- Specialist nurses have enhanced communication skills, by building a rapport with the patient, The patient is encouraged to contact the helpline to expedite admission to the programmed investigation unit, when in severe pain.
- By having excellent communication skills it ensures the patient gets the right treatment at the right time including all the members in the multi-disciplinary team.
- It is essential when communicating with the patient that empathy is conveyed, by being aware of the complex medical history and understanding all aspects of the patients circumstances it can enhance the patient experience, and aid concordance.

Breathing

- The patient does not generally suffer with breathing problems, however due to side effects of the infusion (Flu like symptoms) it is essential that the patient knows the importance of feeling 'well' on the day of the infusion

Eating and Drinking

- The patient has no problems with eating or drinking however can have problems with accessing food and drink in the outpatient setting, the nurses role is to ensure a drink and sandwich is available or obtained, as on occasion the patient attends using hospital transport as the duration of waiting can vary

Eliminating

- The patient has no problems with urinary retention and constipation, however it is essential that the wheelchair bound patient to has easy access to the bathroom, and privacy and dignity is maintained.

Personal cleansing and dressing

- The patient although wheelchair bound can transfer and is independent, as the admission is approximately and hour in duration no special requirements are needed.

Controlling body temperature

- The patient is able to dress independently. advised to wear layers of clothing allowing access to the arms due to the need to cannulate,
- Tympanic temperature is taken on admission to ensure apyrexial before infusion of Zoledronic acid

Mobilising

- Uses electric wheelchair independently when attending the hospital if alone, if attending with husband or other family member has manual wheelchair, when infusing the patient it is important that she can reach all her personal belongings and maintain independence.

Working and Playing

- House and garden adapted to allow maximum independence, has a very full and active life has a teaching qualification but recently stopped due to bone pain, and difficulties with transportation to and from college.
- Need to allow the patient time to talk as a period of adjustment and feelings of self worth.

Expressing sexuality

- Clothes appropriate for the patient,
- Hair care, hair cut is very important as skull involvement to ensure feels feminine
- Wears make up
- Has altered body image therefore it is essential to address psychological needs

Dying

- The patient is not expected to have a greatly reduced lifespan due to the McCune Albright syndrome, presently the pain due to the condition is managed by the use of intravenous bisphosphonates.
- Has previously had Mastectomy for Carcinoma, but has a positive outlook on life.

Specialist Nurses Role and Encompassing the 6Cs

- Specialist nurses are clinical experts in evidence based nursing practice within a speciality and use technical knowledge and insight from patient experience to lead service and redesign it, and improve the quality of care the patient experience through enhanced communication and advocacy skills, By incorporating the 6 Cs this ensures that;
- Care is delivered offering high quality care
- Compassion how care is given through relationships based on empathy, kindness respect and dignity
- Competence Knowledge and skills and to deliver best care on the best available evidence
- Communication no decision about me without me
- Courage to do the right thing, be bold and speak up when things go wrong
- Commitment will make our vision for the person receiving care our professions and our teams happen we commit to take action to achieve this

Conclusion

- Using a holistic model of care for the patient this ensures the patient's needs are met, which is appropriate for their condition. The Roper Logan and Tierney model can be utilised for all conditions and encompasses all aspects of the patient.
- A specialist nurse by having extensive knowledge and management of the disease access to an eminent endocrinology consultant, and an understanding of the systems in place the patient has a seamless episode of care and confidence that the right treatment is given using the best available evidence.



References

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