

FROM CLOWN TO CLONIDINE

Palliative symptom relief – a teamwork



Lilla Erstagården
Children's and youth
hospice



Karin Bäckdahl Pediatrician



1

Pediatric Palliative symptom relief

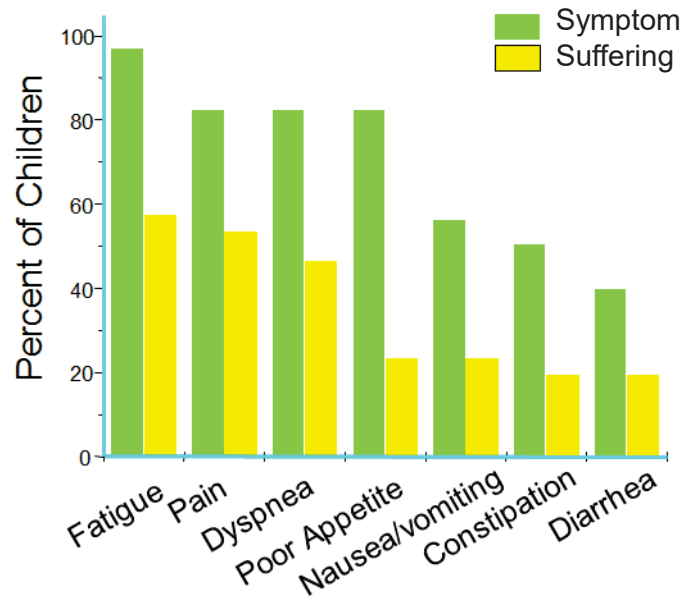
Aim

- Similarities and differences between children and adults
- Similarities and differences between different palliative diagnosis
- How to change the palliative care in different phases through case management



2

Symptom and suffering the last month of life



Wolfe et al. NEJM 2000.

Symptoms and suffering at end of life in children with cancer



3

What we do matters – for a long time

Care related stressors	Experienced	Affected (4-9 yr after)
Pain could not be relieved	46%	57%
Difficult moment of death	32%	57%
Negligent care of my child	46%	41%
Insufficient contact with health care staff after my child's death	43%	33%

Kreicbergs U. et al. J Clin Oncol 2005; 23;(36):9164-71



Palliative Care for Children

TRUST



- Be honest
- Don't promise something you can not keep
- Courage to stay
- Important to include the child as much as possible

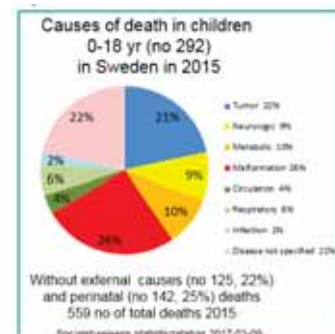
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Groups of patients



- **Neurological diseases**
 - Neurometabolic degenerative diseases - Krabbes
 - Malformation syndroms – trisomi 13, syndroms
- **Oncological diseases**
- **Other**
 - Cardiological diseases
 - Metabolic syndroms
 - Neonatal conditions
 - Respiratory insufficiency



6

Symptom

- Pain
- Fatigue
- Anxiety
- Dyspnea
- Circulation
- Nausea
- Nutrition
- Elimination (Constipation/micturate)
- Skin/Eyes/Itching

- Pain 84 %
- Loss of Appetite 73 %
- Fatigue 63 %
- Nausea/Vomiting 58 %
- Dyspnea 55 %
- Constipation 47 %



7

Symptom relief



Distraction

Clown
och
therpy-
dog



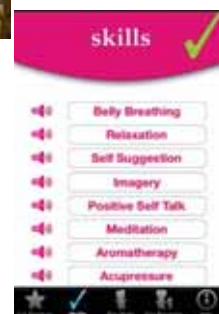
Medicine



Art, music
and play
therapy



Physioterapy



"there's an app for that"



8

Palliative Care for Children

Guidelines for symptom control in children

- There are guidelines for adults – "vårdprogram"
- We are working on guidelines in Sweden to be a part of "vårdprogrammet"
- There are international guidelines –

<http://www.rainbows.co.uk/wp-content/uploads/2011/06/Rainbows-Hospice-Basic-Symptom-Control-In-Paediatric-Palliative-Care-8th-Ed-2011-protected.pdf>

<http://www.starship.org.nz/for-health-professionals/national-paediatric-palliative-care-clinical-guidelines/#All>



9

Pharmacological treatment -routes of administration

- Peroral/gastrostomi/nasogastric tube
- Rectal (younger children)
- Buccal
- Nasalt
- Inhalation
- Cutaneous
- Intravenously /PCA-pump - SVP
- Subcutaneous
- Intratecal
- Intramuscular



10

ANNA

Pediatric palliative care and symptom relief

- Anna was born healthy, but from 2 yrs of age she gradually loses abilities like walk, talk and eat/swallow.
- **Diagnosed with a leukodystrophy - a progressive neuro-metabolic disease without cure or treatment**
- She develops myoclonus and severe spasticity
- She receives gastrostomy for all her food and medicines
- Anna is blind, she cannot talk and its not sure if she can hear



ANNA

Pediatric palliative care and symptom relief

- A large multi-professional team is working with Anna and her family
 - Physical therapy, habilitation, social worker, neurologist
 - *Respite care at Lilla Erstagården*
- Different palliative drugs in high doses against spasticity and pain
 - Baclofen, benzodiazepines and Clonidine (Clonidine also for pain and anxiety)
 - Against presumed neuropathic pain--Neurontin/Gabapentin
 - Sleep regulation with Melatonin/Theralen/Clonidine



Integrated palliative care for a long period of time



Who can be admitted to Lilla Erstgården?

- End of life – prio 1 patients
- Patients in a palliative definition
 - **Respite care**, shorter periods
 - **Post-care** if need of a prolongation of hospital care if acute illness, operation.



No longtime solution of care/respice



13

Care planning at Lilla Erstgården

The care team visits the family in the acute ward/hospital for presentation to the family and to make a care planning

The family is offered to visit Lilla Erstgården children's and youth hospice

Detailed inventory of the patients needs

- Med. tech/ medical needs
- Medical aid
- Nutrition
- Medicines
- Caring needs
- Family needs; parents, sibling & other relatives

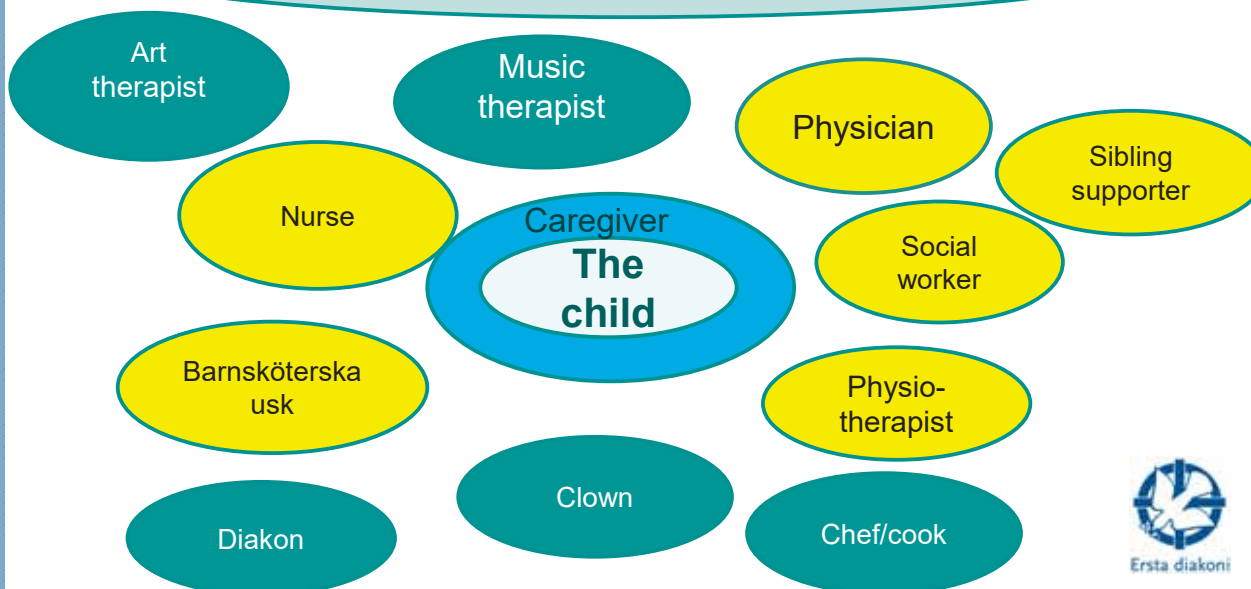


Admittance for end of life care or a respite/post care period

14

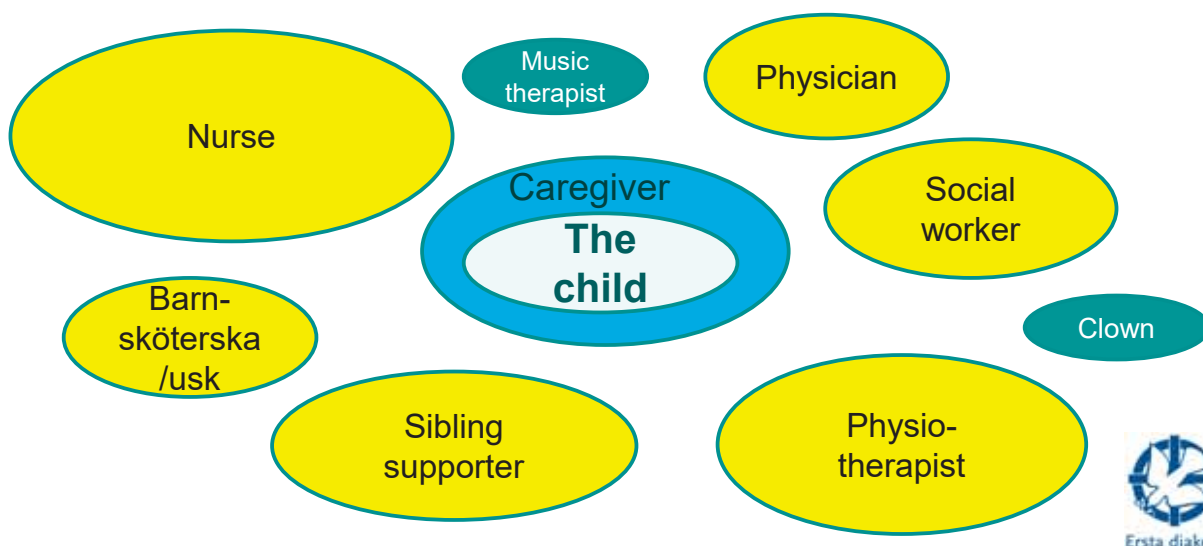
Multi-professionell teamwork at Lilla Erstagården

Hemsjukhus, hemsjukvård (spec.läk, spec.ssk, dietist, fysioterapeuter), habilitering, smärtenhet



Multi-professionell teamwork at Lilla Erstagården

Hemsjukhus, hemsjukvård (spec.läk, spec.ssk, dietist, fysioterapeuter), habilitering, smärtenhet



Pediatric palliative pain

- Is the child in pain? – How do we know?
- What kind of pain?
Nociceptive, Visceral, Neuropathic, Existential?
- Long-term pain compared to acute onset of pain

17



Children with neurological disease/symptoms

- no verbal communication, impaired cognitive ability
- more than one disabilities as EP, dystonia/spasticity, constipation
- All neurological symptoms may be exacerbated due to discomfort, infection, constipation, disturbed sleep and medicine side effects

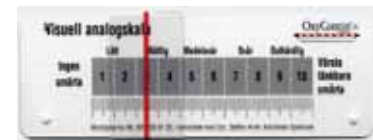
**Often a complex pain situation
must weigh relief against sedation**

**IF WE THINK IT MIGHT HURT
IT PROBABLY DOES!!!**



Pain assessment tools

- **ALPS 1** Newborn - 1 month
- **ALPS 2** 1 month - 3 yr
- **Face scale** From about 3 yr
- **VAS** From about 6 yr
- **BOSS/FLACC** Children non-verbal, neurological impaired



19

rFLACC

Face, Legs, Activity, Cry, Consolability
A Preverbal (Non-verbal) Patient Pain Scale

The Revised FLACC Scale (rFLACC)				
rFLACC	0	1	2	Individual descriptors
FACE	No particular expression or smile	Occasional grimace/frown; withdrawn or disinterested; appears sad or worried	Consistent grimace or frown; frequent/constant quivering chin, clenched jaw; distressed-looking face; expression of fright or pain	'Pouty' lip; clenched, grinding teeth; eyebrows furrowed; stressed looking; stern face; eyes wide open - looks surprised; blank expression; nonexpressive
LEGS	Normal position or relaxed; usual tone & motion to limbs	Uneasy, restless, tense; occasional tremors	Kicking, or legs drawn up; marked increase in spasticity; constant tremors or jerking	Legs and arms drawn to center of body; clonus in left leg with pain; very tense and still; legs tremble
ACTIVITY	Lying quietly, normal position, moves easily; regular, rhythmic respirations	Squirming, shifting back and forth, tense or guarded movements; restlessly agitated (e.g. head back and forth, aggression); shallow, spitting respirations, intermittent sighs	Anched, rigid or jerking; severe agitation; head banging; elevating head; rigid; teeth holding; gasping or sharp intake of breaths; severe spitting	Grabs at site of pain; nods head; clenches fists; draws up arms; arches neck; arms stiffen; turns side to side; head shaking; points to where it hurts; clenches fist to face; hits self; slapping; tense; guarded posturing; thrashes arms; bites palm of hand; holds breath
CRY	No cry/verbalization	Moans or whimpers; occasional complaint; occasional verbal outburst or grunt	Crying steadily, screams or sobs, frequent complaints; repeated outbursts, constant grunting	States 'I'm okay' or 'All done'; mouth wide open & screaming; states 'Owie' or 'No'; gasping, screaming; grunts or short responses; whining, whimpering, wailing, shouting; asks for medicine; crying is rare
CONSOLABILITY	Content and relaxed	Reassured by occasional touching, hugging or being talked to, distractible	Difficult to console or comfort; pushing away caregiver; resisting care or comfort measures	Responds to cuddling, holding, parent stroking, kissing, distant and unresponsive when in pain

(Voepel-Lewis, Merkel, Tait, Trzcinka & Malviya, 2002)



20

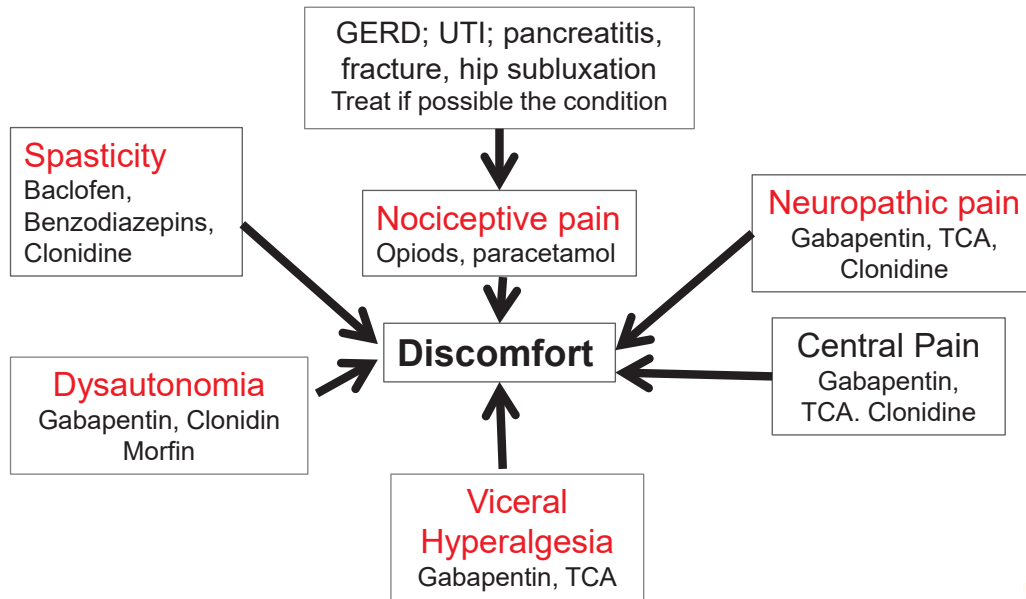
- Non-Communicating Children's Pain List" et al Lynn Breau



- Don't forget one of our most important tools
 - the experience of the parents
 - the experience of the nursing staff
- Together with the child self -assessment and/or a validated assessment tool



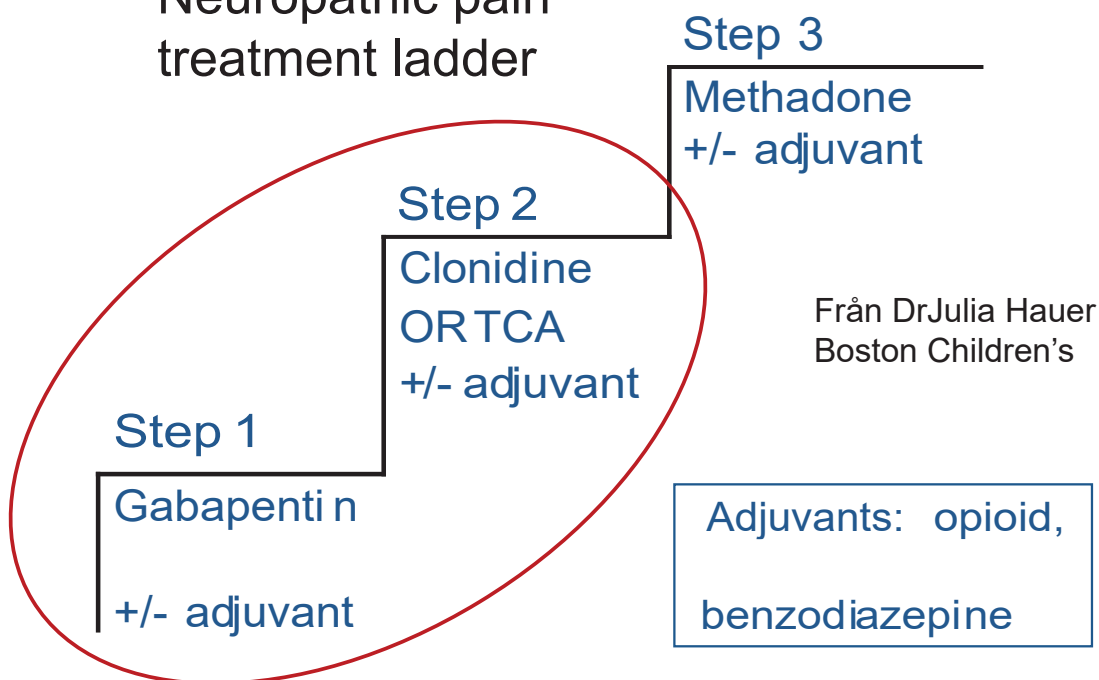
Discomfort in children with severe neurological impairment



23

Neuropathic pain

Neuropathic pain treatment ladder



24

Neuropathic pain

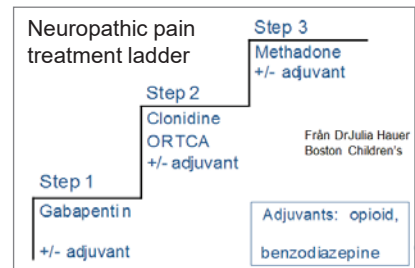
- **Gabapentin** (antiepileptic),

30-45- ? mg/kg/day

Divided by 3 dos/day, increase gradually

less side effects in children than adults?

Pregabalin (Lyrica) – older children?



- **Amitriptyline** (tricycles antidepressant)

1 dose at night

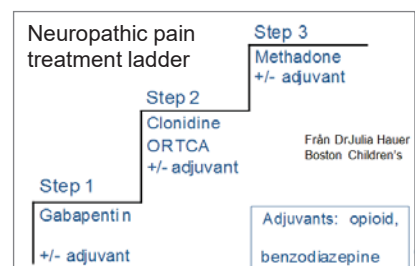


25

Neuropathic pain

Clonidine (Catapresan)

- α_2 -receptor agonist
- oral- tablets/oral solution
- rectally - injection solution/ oral solution
- intravenous injection,
- continuous intravenous infusion with bolus doses
- patch?

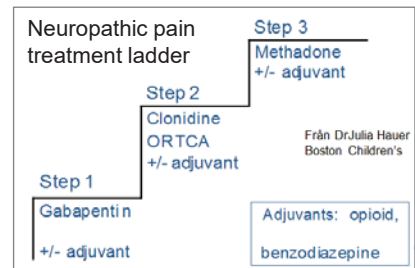


26

Neuropathic pain

Clonidine (Catapresan)

- Effect on
 - pain (neuropathic and nociceptive)
 - spasticity
 - anxiety
- do not affect respiration
- Limited side effects - sedation/hypotension/headache?
- Dose initially 1-3 mcg/kg x 3-4, can be increased
- Large therapeutic window

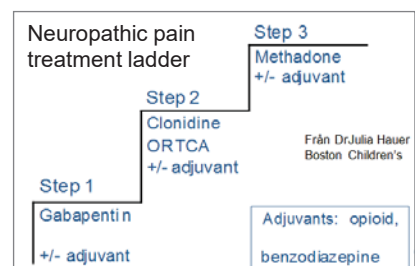


27

Neuropathic pain

Methadone

- NMDA receptor antagonist
- Often a combination of methadone and other opioid
- Methadone according to FASS contraindicated in children



Kontraindikationer

Andningsdepression.
 Överkänslighet mot den aktiva substansen eller mot något hjälpämne.
 Akut obstruktiv luftvägssjukdom.
 Samtidig administrering av MAO-hämmare eller administrering inom två veckor efter avslutad behandling med MAO-hämmare.
 Kontraindicerat till barn.

FASS



Consult someone with experience in children

28

Non-Pharmacological treatment

Integrative methods



- Breathing exercises
- Mucus mobilization
- Massage/ warm/heat/bath
- Physical activity
- Positioning
- Self regulation skills
- Distraction/diversion



29

ANNA Pediatric palliative care and symptom relief

Different palliative drugs in high doses against spasticity and pain

- Baclofen, benzodiazepines and Clonidine
(Clonidine also for pain and anxiety)
- Against presumed neuropathic pain--Neurontin/Gabapentin
- Sleep regulation with Melatonin/Theralen/Clonidine

Gradually worsened


One day intense vomiting with blood and
symptoms of bleeding from the stomach

Pre-planned plan of action



30

Limitations in care

Ställningstagandet innebär:	Document on life-sustaining treatment
<input type="checkbox"/> Ingen begränsning av (livsuppehållande) behandling <input checked="" type="checkbox"/> Avstå från att påbörja livsuppehållande behandling, nämligen: <input type="checkbox"/> Avbryta pågående livsuppehållande behandling, nämligen:	<p>Limitation in care can be a tool to reach the goal of care</p> 
<div> <input type="checkbox"/> MIG (mobila intensivvårdsgruppen) <input type="checkbox"/> Andningsoxygän </div> <div> <input checked="" type="checkbox"/> IVA (intensivvård) <input type="checkbox"/> Vasoaktiva läkemedel </div> <div> <input type="checkbox"/> HIA (hjärtintensivvård) <input checked="" type="checkbox"/> Antibiotika </div> <div> <input checked="" type="checkbox"/> Återupplivning efter hjärtstopp <input type="checkbox"/> Cytostatika </div> <div> <input type="checkbox"/> Pacemaker <input type="checkbox"/> ICD (intern defibrillator) <input type="checkbox"/> Nutrition </div> <div> <input checked="" type="checkbox"/> Invasiv ventilatorbehandling <input type="checkbox"/> Vätska </div> <div> <input checked="" type="checkbox"/> Noninvasiv ventilatorbehandling <input type="checkbox"/> </div> <div> <input type="checkbox"/> Dialys <input type="checkbox"/> </div> <div> <input checked="" type="checkbox"/> Operation <input type="checkbox"/> </div> <div> <input type="checkbox"/> Strålbehandling <input type="checkbox"/> </div> <div> <input type="checkbox"/> Blodtransfusion <input type="checkbox"/> </div>	

OPTIFLOW/ AIRVO?

31

Limitations in care

The medical record should contain

- a clear references to notes where limitation in care has been discussed
- any decisions that have been made by the responsible doctor and patient/family

Documentation regarding life supporting actions shall be clear and easily accessible



End of life Care Plan



Allowing a natural death (AND) and providing palliative care does not indicate a withdrawal of care, but the provision of symptom management; psychosocial and spiritual support and comfort during the end of life period.

The following goals of care were identified:

Symptom management

Important to talk about what we should do to alleviate more than to talk about what we shouldn't do and to document



33

“Ambulance-paper”



- An agreement on the “ambulance-paper” is made between the physician and parents
- Valid for 3 months may be extended if needed
- Documented in the patient's journal in under the keyword “treatment limitations”
- The parents choose whether to use it or not



34

ANNA

Pediatric palliative care and symptom relief

Limitation in care – decisions made in advance

She has an "ambulance-paper" -

Possibility to a direct admission

- Anna is admitted
- No more food/fluids/medications can be given through the gastrostomy
- No intravenous access/no iv fluids or TPN
- What do we do now?
- How can we now alleviate her symptoms?



35

Nutrition

“Äta bör man annars dör man”

- Iv infusion or not? There is a lot of evidence in adult to avoid iv fluid in the end of life we believe it's the same for children.
- What you want to eat – ” if you want to eat”
- Food/fluid in gastrostomy/NG tube or not?
 - When do you stop giving food and fluid?



“Food to give wellbeing and palliation”

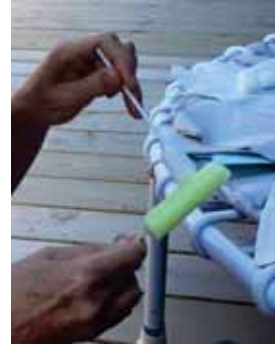


36

ANNA end-of-life care- without needles

No liquid or food could be given

– **Mouth care**



- No medications could be given through the gastrostomy
- No iv or sc access



37

ANNA end-of-life care- without needles

Instead

- **Opioids** - Durogesic patch (0,5 of 12µg/h - About 15-20 mg of po Morfin/day) with rescue dose of Morfin inj solution rectal
- **Clonidine** (inj solution) and **Paracetamol** (supp) rectal x 6 and as needed
- **Midazolam** buccal as needed



38

Patient with severe cancer pain

Opioid ex **Oxynorm 10mg/ml** i PCA pump, infusion with bolusdoses/max1 in 30 min ... or.

When the volume becomes a problem transition into **Hydromorfon 20mg/ml** (7 times more potent opioid)

iv inj of **Clonidine**/ infusion in PCA-pump

Iv opioder and bensodiazepiner as needed

Ketamin infusion (no PCA) **iv in SVP**

Midazolam infusion (PCA-pump)

Propofol infusion (no PCA)



39

Non - opioids



Paracetamol

- antipyretic, analgesic and
- oral/gastrostomy, rectally, intravenously

NSAID

- Antipyretisk och anti-inflammatory effect
- oral/gastrostomy, rectally



40

Around the clock (ATC) or as needed (PRN)?

Important considerations in children who may find it difficult:

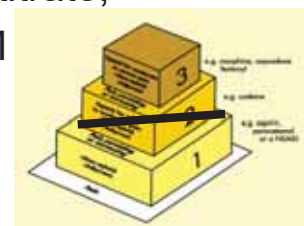
- To describe or define the pain.
- To be believed?
- To dare to tell?
- Or cannot tell – more than maybe when the pain is very strong – ex small children, severe neurological disease children



41

Weak opioids – step 2

- ~~Codeine- is not recommended for children!~~
 - Difficulty with the effect it give, difficult to titrate,
 - advised against in most pediatric pain PM
- ~~Tramadol – we do not use Tramadol~~
 - weak effect on u-receptor
 - Possibly effect on neuropathic pain side effects to



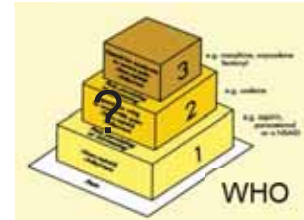
GIVE A "STRONG" OPIOID INSTEAD!



42

α_2 - receptor agonist

Clonidine



An alternative step 2 for children?

- Many routes of administration
- Effect on
 - pain (neuropathic and nociceptive)
 - spasticity
 - anxiety
- Do not affect respiration
- Limited side effects - sedation/hypotension/headache?
- Large therapeutic window



Strong Opioid



text Morphine

- Per oral /gastrostomy at least x4-6/day

0 - 6 mo	0.075 - 0.15 mg/kg/dos
6 - 12 mo	0.15 - 0.2 mg/kg/dos
1 - 5 yr	0.2 - 0.3 mg/kg/dos
5 - 16 yr	0.3 - 0.4 mg/kg/dos
- So many ways of giving opioids.....
- If a regular opioid – always add Naloxone per os
2-12 mcg/kg x 4 prophylactically to prevent
obstipation (older maybe Moventig^R (Naloxegol))



Transdermal fentanyl-patch

- If possible find a dose po/iv first and then transfer to

Transdermal fentanyl in children

- Fentanyl in ethanol gel - Fentanyl Hexal^R
- Fentanyl in matrix - Durogesic^R, Matrifen^R
- Matrix patch-can be divided into smaller pieces for smaller children, but more expensive
- Change recommended after 72 hours – consider replacing after 48 hours in children



45

NMDA receptor antagonist

Methadon

Ketamin

- Continuous low-dose infusion without bolus doses.
- Can be administered orally (bitter) alt given nasally
- Side effects - hallucinations, raises blood pressure



46

ANNA

end-of-life care- without needles

- **Opioids** - Durogesic patch (0,5 12µg/h - About 15-20 mg of po Morphine/day) with rescue dose Morphine inj solution rectal
- **Clonidine** (inj solution) and **Paracetamol** (supp) rectal x 6 and as needed
- **Midazolam** buccal as needed

Doses increases as needed according to symptoms

After 7 days Anna dies calm and alleviated



47

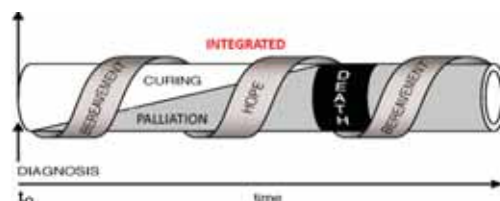
Bereavement support

Home visit by or contact with the social worker

After a month meeting with the team

Thereafter individual support according to the needs of the family

One year of bereavement support



48

What if the child doesn't die?

We have to be prepared to change the care from end of life to more longterm palliation again if needed!

49



How does the team coop?



Reflections over the days work in a staff-meeting at the end of the day

Self care and improvement of care



Structured reflections in a staff-meeting after a death of a patient



Team coaching/guidance/counselling

50



Day of remembrance



A day for families that have lost a child during the past year at Lilla Erstagården



51

Welcome to visit us at Lilla Erstagården



Lilla Erstagården



52