



VENOUS ACCESS IN CYSTIC FIBROSIS

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Introduction

- Today CF is not a pediatric disease with a poor prognosis anymore (half of CF patients are now 18 years older in many countries).
- A success that means new I.V. problematics ..

AL. Stephenson, S. Stanojevic, J. Sykes et al. The changing epidemiology and demography of cystic fibrosis. 2017

L. Regard, C. Martin, P.-R. Burgel et al. Acute and chronic non-pulmonary complications in adults with cystic fibrosis. 2019



Purpose

- To clear on the impact of I.V. therapy and catheterism on CF patients
- To explain how CF patients try to maintain a balance between pleasure and I.V. care's constraints
- To explain the aim of CF center that can be summarized in « Care for life and not a life to care »



CF Definition

- Autosomal recessive genetic disease due to mutations in the gene encoding the CF transmembrane conductance protein (CFTR).
- Multi organic disease (lungs, pancreas, intestine, liver and reproductive organs)
- Newborns are screened and cared for multidisciplinary teams in CF centers
- At least 70 000 CF patients worldwide; more frequent genetic disease in France



CF Definition

ORGAN AFFECTED	TREATMENTS
Lungs: clearance of increased airway secretions decline	Physiotherapy: and inhaled hydrator therapies: daily
Pancreas: exocrine pancreatic insufficiency	Pancreatic enzyme replacement and high-calorie/High fat diet: daily Fat-soluble vitamins (A,D, E, K): daily or less frequently
Higher sweat concentrations of Na and Cl	Prevention++: daily (depends on the place where you live)
Reproductive organs: infertility	Medical assistance to procreate: daily when it occurs
CF-related diabetes (in adults++) and its complications	S/c insulin and monitoring: daily
Constipation	Diet-related measures: daily
Gastroesophageal reflux disease	Diet-related measures and acid blockers: daily
Urinary incontinence (< cough, sneezing)	Physiotherapy, lungs treatment: daily
Sinosonal manifestations (nasal polyposis)	Saline irrigations, topical corticosteroids: daily . Surgery.
Anxiety and depression	Medical treatment and psychologist: sometimes weekly



CF Treatments

ACUTE COMPLICATIONS WITH CF	TREATMENTS
Pulmonary exacerbations	<i>See after</i>
Hyponatremic dehydration	Rehydration with I.V. saline.
Pancreatitis: pain	Food intake rather than fast. Prevention++ (alcohol use): keep in mind
Complications of gallstones (cholethiasis, cholecystitis and cholangitis)	Surgical procedures and I.V. antibiotic courses
Intestinal obstructive syndrom: abdominal pain, vomiting, nausea	Laxative, I.V. hydration, fasting, nasogastric aspiration
Symptomatic nephrolithiasis	Lithotripsy or surgery
CF-related liver disease (cirrhosis)	May lead to liver transplantation if
Malignancies (colorectal or testicular cancer)	Chemotherapy, . . .
Lung transplantation-related complications	<i>But this is another story...</i>



CF and Treatments

In others words:

- CF is here to stay.
- CF daily reminds patient that without daily care, he could less satisfy his ambitions in life.
- CF patient can be fed up with treatments: CF caregivers must keep in mind this for their daily practice



CF Treatment

- Lung disease = major cause of the significant morbidity and mortality
- *Pulmonary exacerbations* and complications (pneumothorax and haemoptysis)
- *I.V. antibiotic* course necessary
- Result: Bacteria eradication or not
- *Repeated ATB courses* can lead to ototoxicity, chronic kidney disease hypersensitivity reactions/allergy.



CF Treatment

- Most of antibiotics are infusable in peripheral veins. No vesicant drug.
- 2 or 3 weeks I.V. course; intermittent or continuous infusions (*attention to drug stability!*)
- For over time, frequency of IV treatments usually increases
- Hospital or homecare (usually by freelance homecare nurses in France)
- Organized by nurse coordinator



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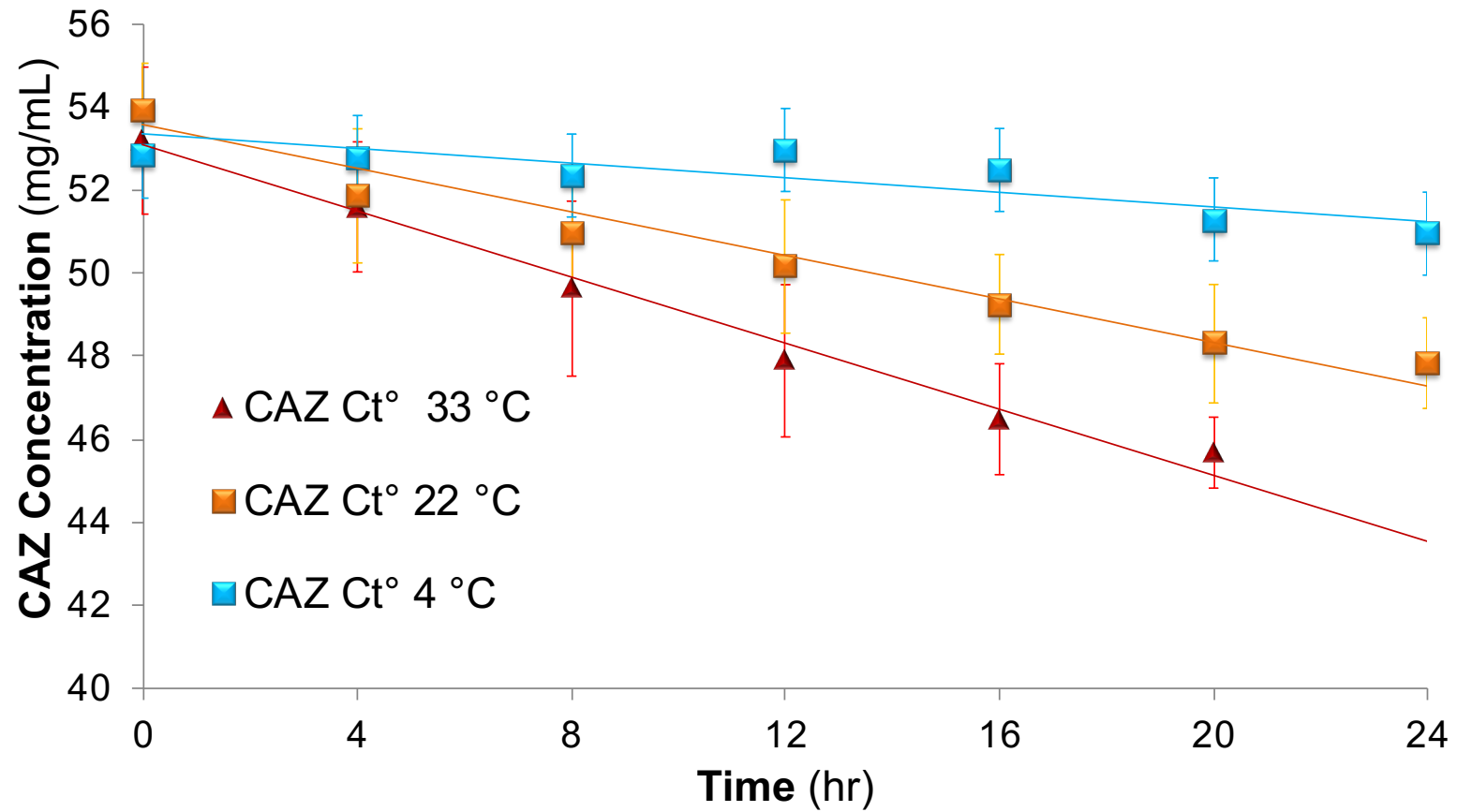


CF treatment

- Antibiotics
- **Ceftazidim** (D. Hubert, R. Garraffo et al . Continuous versus intermittent infusions of ceftazidime for treating exacerbation of cystic fibrosis. 2009).
→ A 24hrs infusion of CAZ is possible and improves CF patients quality of life
- **But ..**

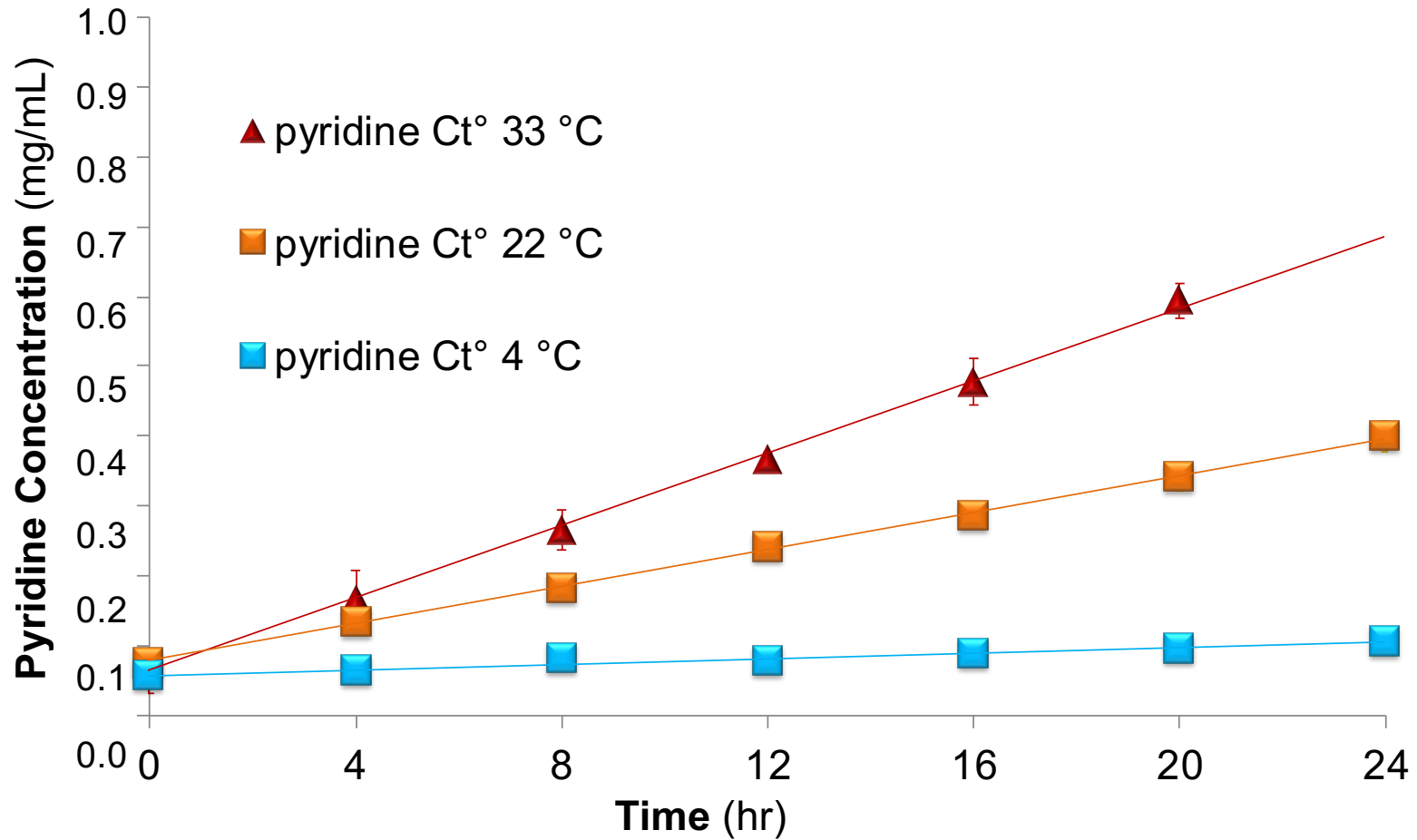


CF treatment





CF treatment





CF Treatment

- Same story with Meropenem: in 2015, 3% of french CF centers prescribed Meropenem over 12 hours
- **Over time, widely use of generics** (SC Schaefer, PA Bison, R Rangoonwala, CJ Kirkpatrick, HA Lehr. 0.2 μm in-line filters prevent capillary obstruction by particulate contaminants of generic antibiotic preparation in post ischemic muscle. 2008)



I.V. Catheters choice

- No international specific guidelines
- Except for DIVA, needle phobic patient, ***short peripheral canula is the first choice***
- If ***short peripheral canula*** insertion gets difficult: ***Midline or PICCline***
- **Midline** is not widely used in France. It could protect central veins from injuries related to catheter insertion, and from LMWH use in case of DVT



I.V Catheters choice

- Problem with Midline and PICC line: no insertion possible in patient home (transport); possible delay between prescription and insertion

- How many PICC can be inserted in a same patient?

(R. Y. Yang, R. Moineddin, B. Connoly. Increased complexity and complications associated with multiple peripherally inserted central catheter insertion in children: the tip of the iceberg. 2012. R. Gnannt, N. Waespe, B. Connoly et al. Increased risk of symptomatic upper-extremity venous thrombosis with multiple peripherally inserted central catheter insertions in pediatric patients. 2018)



I.V. catheters choice

- **PORT** is the best indication in case of the higher frequency of I.V. courses
- It's implanted, ready to use, comfortable when a needle is not inserted
- **But** CF patients and caregivers are not robots but human beings..



Port meanings

- Spc, Midline and PICC are removed at the end of the I.V. treatment, not Port
- Port is proposed when lungs function declines
- Port stays under the skin : CF gets visible (poor experience of brachial port)



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Port meanings

- Port leads to explanations about your health
- Port can't be removed by yourself
- Ports complications can't be compared with peripheral catheter or PICC's complications



Port meanings

- Port 's proposal is a very important step for CF patient and CF team
- It takes time
- Communication between caregivers and patient is key
- Caregivers acceptance/expertise/professionalism are another key



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Homecare

- Prevention of social isolation and infections related to admission in hospital
- Decrease costs for National Health Insurance
- **But homecare must be accepted by patient**
- *They must be organized* (think about the worst cases to provide the better service)
- *Skills and practices of a multidisciplinary team* are required
- Follow-up and assessment are key
- Self-management of I.V. administration/flushing is not so unusual



Complications

- Lumen occlusion > MARSI > venous stenosis > DVT > infection > mechanical complications



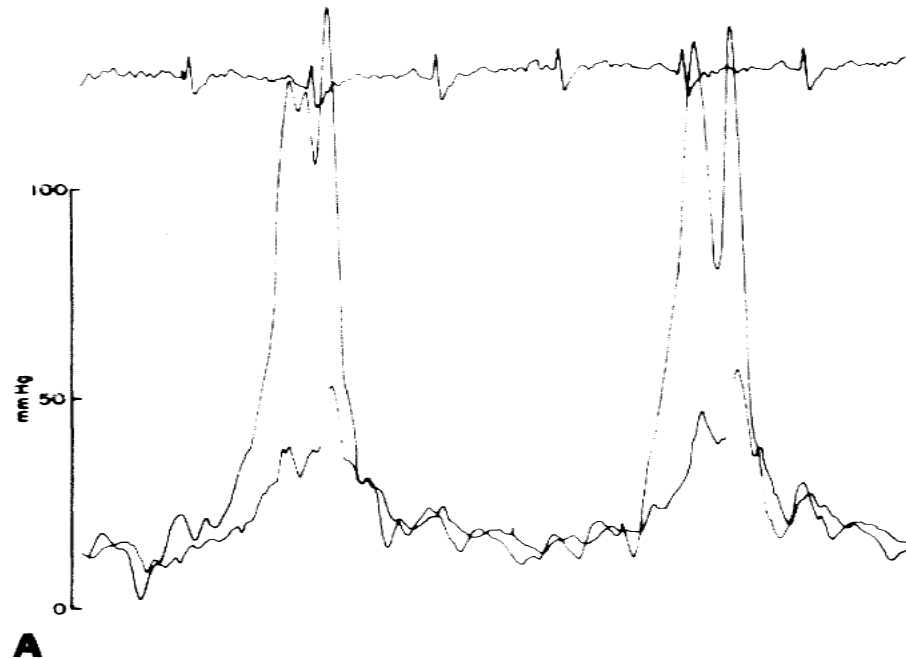
« No blood return. The flow rate got slow. So we still used it »

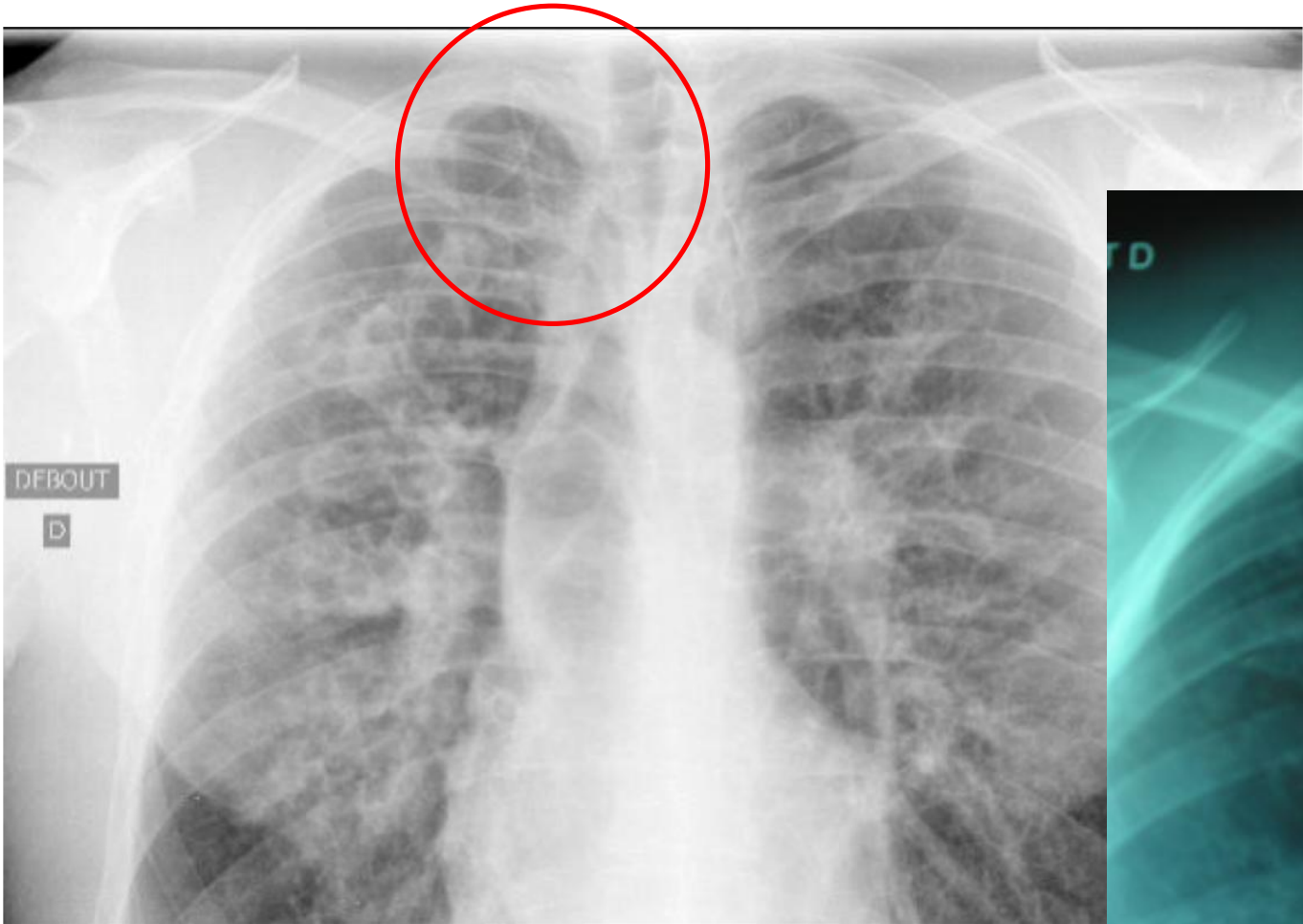


Complications

- Lumen occlusion > MARSI > venous stenosis > DVT > infection > mechanical complications
- **Lumen occlusion** (18% for PICC) caused mostly by cough, improper flushing or/and locking, drugs precipitates

Pressure in Jugular vein during CF patient coughing





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D. Roblin, J.C. Porter et al. Spontaneous migration of totally implanted venous catheter systems from subclavian into jugular veins. Thorax. 1994
W.R. Jacobs, M.H. Zaroukian. Coughing and central venous catheter dislodgement. J Parenteral Enteral Nutr. 1991



Complications

- *Lumen occlusion's treatment:* Fibrinolytic protocol (10 000 UI/mL Urokinase solution)
- *Prevention:* training about proper use of drugs and catheter; elastomeric pump and volumetric pump use; Saline lock.

G.A. Goosens, Flushing and locking of venous catheters: available evidence and evidence deficit. 2015

Ming-Shen Dai. Catastrophic heparin-induced thrombocytopenia/thrombosis syndrome related to the use of a Port-A-Cath in a breast cancer patient receiving chemotherapy. 2004



Complications

- **MARSI.** Under-estimated problem. Several causes: unproper use of DM or antiseptic, dressing, antiseptic's allergy or intolerance, antibiotic effects on skin microbiota?
- *Prevention:* knowledge about MD and antiseptic, monitoring, patients follow up
- *Treatment:* no systematic solutions



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Complications

- **Venous stenosis and DVT**
- Venous stenosis: you can't insert a catheter anymore.
- Question: how many catheters can you insert in the same patient?

(RY Yang, B. Conolly et al. Increased complexity and complications associated with multiple peripherally inserted central catheter insertions in children: the tip of the iceberg. 2012)



Complications

Symptomatic DVT related to PICC (8%) and Port (16%)



B. Lipuma



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C. Dupont, P.-R. Burgel et al. Complications of peripherally inserted central catheters in adults with cystic fibrosis or bronchiectasis. JVA 2015
L. Moira, R. Tonelli et al. Complications of indwelling catheters in Cystic Fibrosis. Chest. 2000



Complications

- *DVT Risk factors:* catheter diameter, multiple lumens, poor nutritional status, Burkholderia cepacia infection, several PICC yet inserted, thrombophilia related to CF

(Barker-Hermann. Prevalence of thrombophilia and catheter related thrombosis in CF. 2005; D. Serisier. Catheter related thrombosis associated with elevated factor VIII levels in CF. 2006; A. Munck Central venous thrombosis and thrombophilia in CF: a prospective study. 2014; T. May. Complications of long and intermediate term venous catheter in CF patients. 2017).

- *Treatment:* No CF-specific recommendations for the management of DVT in CF patients. LMWH and vitamin K antagonists are used. Duration of atcg therapy must be discussed on a case-by-case (**high risk of hemoptysis**)



Complications

- **Infection** rates remains low : incidence of 0% (for PICC) – 10.3% (for Port)
- Most common organism cultured from the bloodstream: Fungi (Candida species++) < exposure to antibiotics (Withers. Fungal septicemia in patients with CF associated with TIVAD. 2007 ; C Mc Carthy, O. O'Carroll. Risk factors for TIVAD –associated complications in CF. 2017)
- *Prevention and treatment* follow the published guidelines



Complications

- **Mechanical complications (6,8%)** L. Moira, R. Tonelli et al. Complications of indwelling catheters in Cystic Fibrosis. 2000

- Origin: improper use and de-occlusion



A 16 years old Port a Cath



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Port removal

- When the port should be renewed? Should be systematically renewed?

Frequency of port replacement: 3 years (3%) to 5 years (9%) and « as long as functional (47%)



CONCLUSION





What did CF learn us about venous access?

- Patients get older → number of veins is limited → we must protect them
- The « Golden Cath. Quartet »: Patient, treatment, vein, catheter must be taken into account by caregivers.

But a « Golden Cath. Quintet » is really a better idea.

- Research is going on
- Caregivers education and training are key
- Relationships between patient and CF team is fundamental