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Preventing and treating cancers in people with intellectual disability-

An interview with Dr Daniel Satge, one of the founders of the ONCODEFI project in France.
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Hello Daniel, Please tell us about the specialty you have been working in?

I am a pathologist. I have been diagnosing diseases using a microscope for the past 31 years! In autumn 2011 I gave this up to work full time preparing for the ONCODEFI project.

You have a background in Cancer pathology? How did you become interested in working in the field of intellectual disability/Down syndrome?

I started my career in a cancer hospital in Strasbourg. I then practised as a general pathologist in Tulle's hospital for 23 years. This work was mainly focused on cancer. A MD thesis on foetal and neonatal cancer made me aware of the link between genetic diseases and cancer, and I was fascinated that so little was known about this area. For example, one such link is Down syndrome and neonatal leukaemia. I closely followed the medical literature on Down syndrome and neoplasms from that time. My interest in tumours in other intellectual disabilities (ID) came naturally; first as a comparative population with Down syndrome, and subsequently to explore the link between ID of any cause, genetic or not, and cancer.

Who are the lead professionals involved in the care of people with

ID in France? How do the training and service provision in this field work in France?

In France, lead professionals involved in the care of people with ID are mainly psychiatrists and to a lesser extent paediatricians and/or neurologists. Dr Bernard Azéma who is ONCODEFI's psychiatrist, and who is at the root of this project, has worked all his professional life in the field of ID. He is a recognized specialist in ID, particularly in autism, in France. In France, psychiatrists receive training in ID as part of a broad training programme, but there is no formal separate training as a sub-specialty degree and the availability of specialist services is limited. To my knowledge there is no in-patient facilities specifically for people with ID and generally people with ID are treated in mainstream child or adult services.

Coming back to your area of expertise, please tell us briefly why you think this (cancer in people with ID) is an important clinical issue?

How does this overlap with other important issues in ID (e.g. autism, challenging behaviour, genetic syndrome, dementia and epilepsy)? According to recent epidemiological incidence data, cancers are globally as frequent in people with ID as they are in the general population. For instance, in France we expect to see 7,000 new cancers among people with ID and in the world around 700,000 cases annually. Second, cancers in people with ID are different from the general population in terms of their distribution, age at onset, and especially clinical presentation. Third, the association between cancer and ID is not well understood, e.g. conditions such as autism and genetic syndromes have a particular tumour profile and need specific monitoring techniques and in some cases treatment modifications. Unfortunately, the historic belief that cancer is rare in this population is still prevailing. The consequence is that cancer is

often diagnosed at an advanced stage and might be presented or accompanied with signs and symptoms of challenging behaviour or mental illness. This reduces the chance of a cure and increases the cost/burden of treatment (emotional, familial, and psychological as well as financial) for professional and families alike.

Daniel, please tell us about your interesting project, ONCODEFI. What is it exactly?

The ONCODEFI project aims to provide the best cancer care for people with ID. It includes four therapeutic teams, a documentation centre with a website and a research core. The project was set up by an oncologist (Pr Stéphane Culine), an ID psychiatrist, who worked with the European team POMONA for health indicators of people with ID (Dr Bernard Azéma), and a pathologist (myself).

Was there any evidence based research work behind its conceptualisation?

There is not much literature on cancer and ID compared with the data necessary for optimal adapted care. We now know from epidemiological studies that cancer is frequent among people with ID. We know from various sources that some organs such as those of the digestive tract are more at risk of developing a malignancy. Other organs such as the breast have a similar cancer risk as women in the general population, but organs such as the lung have a reduced cancer risk compared with the general population. There is an important gap in knowledge with regard to the features of tumours at diagnosis and adapted treatment. Apart from significant work from some teams, mainly in UK, on screening, access, disclosing-information and palliative care, so much more remains to be studied and researched.

Could you please name some important literature published in this area?

I would suggest a 2001 publication from the British Institute of Learning Disabilities (BILD) entitled "Cancer and people with learning disabilities" by James Hogg and colleagues, which is a good introduction to the subject area. Irene Tuffrey-Wijne recently reported on the delay in cancer diagnosis and consequences for people with ID in her very interesting book "Living with learning disabilities, dying with cancer thirteen personal stories" (Jessica Kingsley Publishers 2009). We also contributed to the subject with a book chapter "Cancer in persons with intellectual disability, current data" in the book "Mental retardation research focus", (Nova Science Publishers New York 2007). In a more recent book, "Cancer in children and adults with intellectual disabilities" (Eds. D Satgé and J Merrick), some important issues for the cancer care of patients with ID are addressed (Nova Science Publishers New York 2011).

Who is the sponsor for ONCODEFI?

This autumn, ONCODEFI received a 450,000 euros grant from the French National Cancer Institute (INCa) for interventional research over a three-year period. We are now ready for the clinical component of the project which includes a paediatric oncology team and three adult oncology teams in Montpellier; this component will start as soon as we receive financial support. We are currently looking for funders.

What are the challenges of this project?

We face a number of challenges in this project. First, we need to make people aware of the double burden of cancer and ID. Second, the project involves the joint-working of health professionals in two different areas, cancer and ID; it is important that they work together effectively. Currently our main challenge is to raise funds to allow the clinical component of the project to begin.

What are the main aims of the project? Are there any targets, short term and long term?

The primary aim of ONCODEFI is to provide the best adapted care for people with ID for prevention, screening, diagnosis and treatment of cancer. For this, we need to gather and synthesize all information and data on the subject and conduct necessary epidemiological and clinical studies.

Our short-term targets are to begin clinical work on children and adults, develop the website and conduct research for evaluation of current aspects of tumours at diagnosis. Our long-term targets are to develop guidance, provide information to all the French regions, and to include ONCODEFI in the international network of ID and cancer. We are currently preparing the research work and the documentation centre website which will go live in January 2013.

Who are the main professionals involved in the project?

In addition to the three professionals mentioned above (S Culine, B Azéma, D Satgé), the main professionals involved in ONCODEFI project are the head of teams: Professor Nicolas Sirvent (pediatric oncology), Professor Marc Ychou (medical oncology), Doctor Xavier Rébillard (urology surgeon), Doctor William Jacot (medical oncology) who work in the university hospital in Montpellier, in the anticancer hospital Val d'Aureille and the Clinique de la Mutualité. We are also linked to the University Clinical Research Institute (IURC) (Director Professor Jean-Pierre Daurès) and the CREA-ORS-Languedoc Roussillon (Director Mr François Clerget) for some aspects of the research project, particularly epidemiology, public health and psychology research.

Is this a local or national project?

Although localized in the south of France, ONCODEFI is truly a national project with the aim of providing information on cancer in ID to all regions of France and creating a network for the best care of cancer in people with ID. Additionally, we will be happy to learn experiences from other teams who treat patients with ID and cancer abroad, and we will be happy to share our experiences in Montpellier.

Do you know of any similar projects/centres in countries other than France? How can other professionals or centres abroad collaborate with the project? Is there any plan to collaborate with other centres abroad?

Teams in UK and in Norway especially, have conducted important research on cancer and ID. However, I do not know of any similar projects to ONCODEFI (treatment, research, information) which involve gathering all data, building a website and conducting epidemiological, psychological and oncological research specifically focused on cancer in people with ID. We would be very pleased and interested to collaborate on the

various aspects of cancer and ID with interested teams in other parts of the world. I can be contacted at: daniel.satge@oncodefi.org . Dr Bernard Azéma can also be contacted at bernard.azema@oncodefi.org

Finally, what would be the take home message, especially for ID professionals working in the UK?

I would like to stress that cancer is frequent among children and adults with intellectual disability. Awareness is paramount because the earlier the cancer diagnosis, the better the prognosis and consequent reduction in the suffering of patients and families, carer burden and the burden on oncological teams. For more information, readers are encouraged to go to our current website www.oncodefi.org which provides information on the project in French as well as English.

Merci beaucoup Daniel, pour nous offrir l'occasion de parler avec vous de ce projet très brillant!

Je vous en prie, Reza! Merci à vous aussi!

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