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TITLE OF CASE ***Do not include "a case report"***

Management of patients with learning difficulties and testicular cancer; overcoming barriers to improve care.

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SUMMARY ***Up to 150 words summarising the case presentation and outcome (this will be freely available online)***

There are significantly poorer cancer-specific survival rates in patients with learning difficulties (LD) including for testicular cancer. Issues such as delayed diagnosis, compliance, consent and needs for individualised care and support needs can make it more demanding to manage these patients in a busy clinical environment. Our case highlights a patient with learning disabilities who presented with advanced testicular cancer due to delayed detection, and highlights the challenges needing consideration to ensure such patients receive good care. This report focuses on the This report highlights an example of adapting curative intent treatment to an individual with complex needs, and the importance of patient and family involvement in decision making to ensure these vulnerable patients receive safe and effective healthcare. The practicalities of multidisciplinary team working in the context of key legislation and existing frameworks to guide practice in the management of LD patients are also discussed.

BACKGROUND ***Why you think this case is important – why did you write it up?***

People with learning disabilities (LD) account for approximately 2.5% of the general population in the United Kingdom. People with LD were previously associated with significant mortality at a young age, but improving health and social care for this population means that they are increasingly being diagnosed with other conditions, including cancer. Testicular cancer is the most curable of the solid cancers. However, the cure rates vary largely depending on prognostic group and tolerability of intensive chemotherapy treatment and there is evidence to suggest these patients have greater mortality than patients without LD. Detection of cancer during the early stages is a potential factor that may influence outcomes. People with LD may not only present differently but may also have different treatment needs from the general population. These range from the amount and form of social support, capacity for consent to treatment, and consideration of treatment options based on a balance of risks and benefits. Our case highlights a gentleman with LD, who presented with advanced testicular cancer due to delayed detection, and highlights the challenges and steps needing consideration to ensure such patients receive the best care possible from the multi-disciplinary team.

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CASE PRESENTATION ***Presenting features, medical/social/family history***

A 61-year-old gentleman presented via the Accident and Emergency Department with a large inguinal and testicular mass. It was not possible to glean from the patient how long it had been present, and was detected by his sister who attended Accident and Emergency with the patient. He had a background of learning difficulties. He had a high level of pre-morbid independence and lived alone in a flat. He could previously mobilise independently and go out to do his own shopping. His sister acts as his main carer and visits him almost daily. His medical history included diabetes and hypertension, and his drug history included risperidone, tamsulosin, metformin, simvastatin and felodipine. He was a non smoker and did not drink alcohol

On clinical examination, he was thin and found to have a solid mass in the right inguinal region measuring more than 15cm in diameter. His right hemi-scrotum was also filled with a mass that appeared to encompass the entire testicle. His Eastern Cooperative Oncology Group (ECOG) performance status was 3.

INVESTIGATIONS ***If relevant***

His tumour markers were performed and were as follows: alphafetoprotein (AFP) 2.1 (normal range,NR 0-5.8), human chorionic gonadotrophin (hCG) 420 (NR 0-2), lactate dehydrogenase (LDH) 4618 (NR 240-480). His full blood count was within normal range, and his creatinine was 138 (NR 59-104), with a GFR of 52 mls/min.

The mass was biopsied percutaneously and revealed a pure classical seminoma, without non-seminomatous elements. An ultrasound of the testis and inguinal region revealed multiple heterogenous vascular lobulated soft tissue masses which were similar in appearance to a large right groin nodal mass (**Figure 1**). Full body staging CT scan was performed (**Figure 2),** which revealed involvement of the pelvic nodes. However no evidence of distal visceral metastases was present. This rendered this gentleman in the group of good prognosis metastatic seminoma based on the International Germ Cell Cancer Consensus Group (IGCCCG) criteria (**Table 1**) [8].

DIFFERENTIAL DIAGNOSIS ***If relevant***

Differential diagnoses included inguinal hernia, epididymo-orchitis and trauma.

TREATMENT ***If relevant***

In advance of the clinic consultation, the learning disability nurses from the patient's local hospital wrote to the specialist centre, advising to book for a prolonged consultation considering his learning disabilities. The patient attended the consultation with his sister and niece, and was seen by the consultant and Clinical Nurse Specialist. The diagnosis of metastatic seminoma was explained, and an assessment of capacity was undertaken. Throughout the consultation, it was felt that the patient could understand, and repeat back key points of information, although retention of this information was only for a brief period. The need to treat with curative intent with cytotoxic chemotherapy was explained. Standard treatment with bleomycin, etoposide and

cisplatin (BEP) offered a cure rate of over 90% but requires an inpatient stay of 3-5 days in each 3-week cycle, together with outpatient chemotherapy appointments on day 8 and day 15 of each cycle. There is also a risk of neutropenic sepsis and other well-described toxicities, which to some extent rely on the patient on being able to phonate and express that they are unwell and subsequently contact the medical team for advice and help. Given the patient's renal impairment there was also a potential risk of hospitalisation, deterioration of renal function and ototoxicity, which may have further impaired the patient's quality of life. The second option, was carboplatin at a dose of area-under-the-curve (AUC) of 10 [1]. This has been shown to have a relapse free survival rate of 94%, albeit in a small number of patients treated to date. This is an outpatient regime administered once every 21 days, necessitating no inpatient stay and a very low rate of inpatient admission for complications such as febrile neutropenia [1]. There is also an improved profile with regards to ototoxicity and renal impairment.

Taking his co-morbidities into consideration, a decision was made between the clinicians, patient and family to proceed with three cycles of carboplatin AUC10.

OUTCOME AND FOLLOW-UP

The patient's sister contacted the chemotherapy hotline three times during the cycles of chemotherapy to discuss concerns about new symptoms, none of which required admission or a consultation with a doctor. Prior to each cycle the patient was seen, along with his sister, in a consultant oncologist clinic to confirm that the patient was fit for treatment. The patient tolerated three cycles of outpatient chemotherapy extremely well with minimal side effects- including grade 1 diarrhoea and mild fatigue. Throughout treatment the patient's tumour markers normalised and the right inguinal mass and testicular mass clinically reduced in size.

After completion of three cycles of chemotherapy a CT/PET scan was completed and discussed at a supraregional MDT prior to discussion with the patient and his sister. This demonstrated an excellent size criteria partial response but with low grade FDG metabolic activity in the right inguinal region, but no new sites of disease or evidence of metabolically active disease elsewhere. Resection of residual disease with a right orchidectomy and right inguinal node dissection was advised. This was undertaken and showed only necrotic cancer with no viable tumour seen. The patient remains in remission 8 months later and will be followed up for 10 years.

DISCUSSION ***Include a very brief review of similar published cases***

Although cure rates for testicular cancer are high, it remains the third leading cause of cancer death amongst men aged 18 to 50. European Society of Medical Oncology (ESMO) guidelines suggest that patients with Stage II/III seminoma receive a combination of bleomycin, etoposide, and cisplatin (BEP) chemotherapy over three 21-day cycles if they fall in the good prognosis group, and four 21-day cycles if they fall within the intermediate or poor prognosis groups. However, there is a reasonable burden of toxicity, alongside commitments of weekly treatments

and blood tests over the period of treatment of 9-12 weeks. [Honecker et al 2018, Annals of Oncology 29: 1658-1686. The three weekly Carboplatin AUC10 regimen, based on phase II data, has been proven to be of high efficacy in patients with good prognosis metastatic seminoma [Tookman et al, reference 1] and more recently has been shown to preserve health-related quality of life when compared to combination cisplatin-based chemotherapy [Milic et al, A Qualitative Analysis of the Impact of Carboplatin AUC10 on Physical, Work Functioning and Bone Marrow Toxicity Among Seminoma Patients – A Single-Centre Experience, January-February 2019, 33;233-237]. The selection of the Carboplatin AUC10 regimen was made after careful and extensive discussion with the patient, family and the multi-disciplinary team, and has produced good outcomes for this patient to date.

Up to 1 in 50 people in the UK have a learning disability [Public Health England (2016). Learning disabilities Observatory. People with learning disabilities in England 2015.]. A study by Afshar et al showed that patients with learning disabilities are approximately four times more likely to die of testicular cancer compared with the population at large [2]. Over a period of 14 years (2001-2015), a total of 158138 male patients identified with a LD 331 had testicular cancer, and 32 died of the disease. In the general population, 25675 had testicular cancer, and 713 died of the disease. The 5-year cancer-specific survival rate was 90.8% in the group with learning disabilities and 97.2% in the general population. The 10-year survival rate was 88.4% in the LD group, compared with 96.8% in the general population. This highlights significantly poorer cancer-specific survival rates in the LD population.

This could be attributed to many factors, including delayed diagnosis due to failure to report abnormalities and a lower rate of testicular self-examination after initial diagnosis. Despite the high-level of premorbid independence of our patient, he did not present until the testicular mass was very large and only after being prompted by his sister. NICE guidelines recommend offering patients with LD an annual physical health with their GP, using a standardised template such as the Cardiff health template. This constitutes a holistic assessment of a physical assessment, a medication review and engagement of a close family member of social care worker, and may mitigate some of the problems including late presentation as exhibited by this case.

This study also highlighted individual challenges of obtaining informed consent for patients with LD, which may have affected quality of treatment. Access to clinical trials is also likely to be limited given the restrictions evident in the Mental Capacity Act to safeguard patients with learning difficulties [3]. Here we describe a case of successful treatment of large volume metastatic testicular cancer in a patient with LD treated with an adapted chemotherapy regime and post-chemotherapy surgery.

A qualitative study of 13 patients with LD and a diagnosis of cancer highlighted that participants' cancer experiences were shaped by previous experience of life, including lack of autonomy and power, and a reliance on their carers to make decisions for them. It also noted that clinicians did not frequently assess their capacity, and hence patients depended on others to negotiate

contact with the healthcare system [4]. This could lead to delayed cancer diagnosis and a lack of treatment options being offered.

In the Afshar et al study of people with LD and cancer, a common feedback point was that patients felt disempowered and disengaged from the consenting process and treatment options, despite having the capacity to understand the majority of the information being conveyed [2]. The right of the patient to consent to or refuse treatment is encoded in the Mental Capacity Act of 2005, which specifically emphasises on the right and involvement of the patient even in the presence of a mental health issue. In the case of a LD patient who lacks capacity, the Mental Capacity Act of 2005 advises the presence of a Mental Capacity Advocate Service to help ensure best interest decisions are made [3]. The Disability Discrimination Act 2005, The Carers Act 1995 and the Human Rights Act 1999 are also key legislations, which help to guide health care professionals in the management of LD patients.

In this case the patient was involved in decision-making as much as possible, along with the support of the clinical nurse specialists and close family members. Many patients with LD have different care and support needs from the general population. There has been a drive to acknowledge these different needs since the "Healthcare for All' inquest around the care that LD patients receive in hospital [5]. It was found that health inequalities were present in comparison to the general population and consequently believed to lead to premature deaths in patients with LD. The inquest stated that carer's opinions were often undervalued, and they were expected to provide care beyond their capabilities; which should have been provided by the healthcare system. It found clinical staff had inadequate understanding of key legislation that is vital to help minimise discrimination and health inequalities for LD patients. Another barrier noted included poor recognition within the healthcare system to implement adaptations needed to care for LD patients. Suggestions for these difficulties included lack of undergraduate and current staff training for the caring of LD patients. The "Healthcare for All' inquest has been vital in driving implementation of systems such as the Public Health Observatory [6], confidential inquiries into premature deaths of all people with LD, mandatory training in undergraduate clinical training, improved monitoring of standards, improved data collection of LD patient's pathway through healthcare and encouragement of annual health checks for LD patients.

Acknowledging these issues, certain adaptations can address potential barriers to delivering excellent care. Examples include longer appointment times and flexibility of appointment scheduling-as was utilised in our patient's case. This has been highlighted and encouraged by the charity Mencap, in association with NHS England, with their 'Don't Miss Out' campaign [7]. Other adaptations include; easy to read information, patient passports, individualised management plans, highlighting communication needs on electronic systems and encouragement of carer involvement. An enhanced support network for LD patients is vital and resources available include Mencap charity, mental capacity advocates, LD liaison nurses, community learning disabilities teams and cancer screening liaison nurses.

Another important key point in the care pathway for LD patients with cancer (but equally

applicable to other diagnoses) are end-of-life care discussions and planning. NICE guidelines [Care and support of people growing older with learning disabilities 2018, NG96] as well as the Care Quality Commission (CQC) independently highlight the importance of building a rapport and effective communication with patients with LD as they approach end of life. Communication could be assisted by aids, such as the usage of the Disability Distress Assessment Tool (DisDAT) to facilitate symptom reporting, and should ideally involve a member of the family or a healthcare or social care professional who knows the patient, while not depriving the patient of autonomy. The CQC had also highlighted very successful cases where the learning disability nurse takes on the role of the care coordinator, working closely with healthcare professionals and family.

In summary, our case highlights the importance of clear and close communication between the patient, family and multidisciplinary team both in the primary and secondary care settings to enhance outcomes of patients with LD. Seeing that up to 2% of the population has a learning disability, it is essential that clinicians equip themselves with the additional needs of this subpopulation of patients and the availability of resources and support networks to enhance their care.

LEARNING POINTS/TAKE HOME MESSAGES ***3 to 5 bullet points – this is a required field***

- A growing number of patients with LD are being diagnosed with testicular cancer
- They are at risk of late presentation due to a lower rate of self-detection, and have different care and support needs, and may have other co-morbidities that influence treatment options.
- Involving patients with LD in conversations as much as possible about diagnosis, as well as the benefits and risks of different treatments is recommended.

REFERENCES ***Vancouver style (Was the patient involved in a clinical trial? Please reference related articles)***

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4. Tuffrey-Wijne Irene, Bernal Jane, Hubert Jane, et al. People with Learning Disabilities who have cancer: an ethnographic study. *Br J Gen Practice* 2009, 59: 503-509.

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7. Mencap. Don't miss out. Available at: https://www.mencap.org.uk/press-release/over-three-quarters-people-learning-disability-are-missing-out-vital-support-gp. [Accessed March 2018].

8. Mead GM, Stenning SP. International Germ Cell Consensus Classification: a prognostic factor-based staging system for metastatic germ cell cancers. International Germ Cell Cancer Collaborative Group. *J Clin Oncol* 1997,15: 594-603.

FIGURE/VIDEO CAPTIONS ***figures should NOT be embedded in this document***

Figure 1: Right testicular mass appearance on scrotal ultrasound

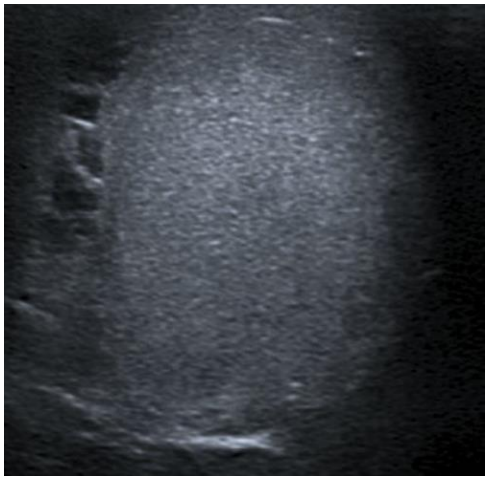

Figure 2: Staging CT scan reveals large inguinal and testicular mass

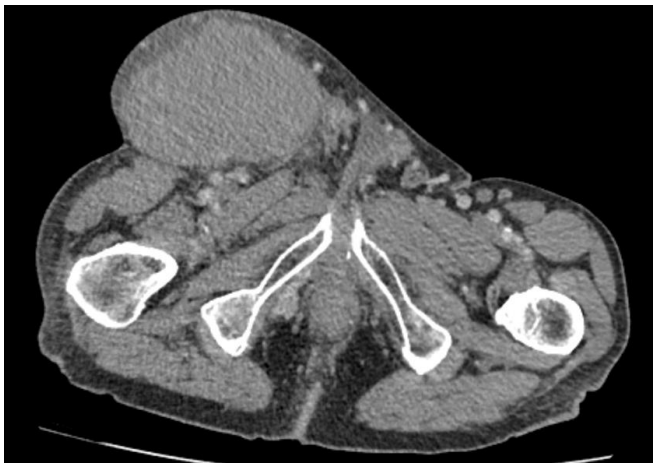

Table 1: IGCCG criteria definition [8].

GOOD PROGNOSIS	
NON-SEMINOMA	**SEMINOMA**
Testis/retroperitoneal primary ***and*** No non-pulmonary visceral metastases ***and*** Good markers - all of *AFP < 1000 ng/ml and* *hCG < 5000 iu/l (1000 ng/ml) and* *LDH < 1.5 x upper limit of normal* **56% of non-seminomas** **5 year PFS 89%** **5 year Survival 92%**	Any primary site **and** No non-pulmonary visceral metastases **and** Normal AFP, any hCG, any LDH **90% of seminomas** **5 year PFS 82%** **5 year Survival 86%**
INTERMEDIATE PROGNOSIS	
NON-SEMINOMA	**SEMINOMA**
Testis/retroperitoneal primary ***and*** No non-pulmonary visceral metastases ***and*** **Intermediate markers - any of:** *AFP ≥ 1000 and ≤ 10,000 ng/mL or* *hCG ≥ 5000 iu/l and ≤ 50,000 iu/l or* *LDH ≥ 1.5 x N and ≤ 10 x N* **28% of non-seminomas** **5 year PFS 75%** **5 year Survival 80%**	Any primary site ***and*** **Non-pulmonary visceral metastases** ***and*** Normal AFP, any hCG, any LDH **10% of seminomas** **5 year PFS 67%** **5 year Survival 72%**
POOR PROGNOSIS	
NON-SEMINOMA	**SEMINOMA**
Mediastinal primary ***or*** **Non-pulmonary visceral metastases** ***or*** **Poor markers - any of:** *AFP > 10,000 ng/ml or* *hCG > 50,000 iu/l (10000 ng/ml) or* *LDH > 10 x upper limit of normal* **16% of non-seminomas** **5 year PFS 41%** **5 year Survival 48%**	**No patients classified as poor prognosis**

PATIENT'S PERSPECTIVE ***Optional but strongly encouraged – this has to be written by the patient or next of kin***

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