# Developing an Integrated, Multidisciplinary Clinic for Duchenne Muscular Dystrophy Patients and their Carers from across the Wessex Region.

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Duchenne Muscular Dystrophy (DMD) is an X-linked recessive neuromuscular condition characterized by muscular wasting, and a progressive dilated cardiomyopathy.

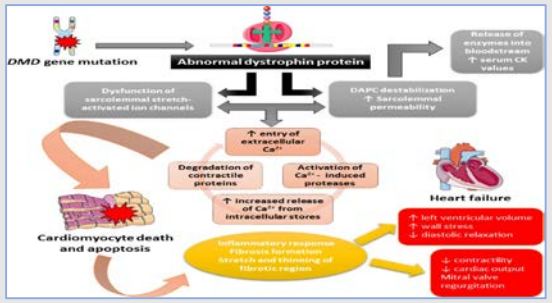


Figure 1. Pathophysiology of cardiac dysfunction in DMD[1]

DMD affects around 1:5000 male live births. Diagnosis is typically made in early childhood but lifelong, ongoing medical assessment and treatment are required to slow the progress of muscle fibre degeneration, support ventilatory function, and offer cardioprotective medical therapy.

## Aims of the Project

1. **Improve the experience of transition to adult care for DMD patients and their carers.**
2. **Improve MDT communication between healthcare professionals caring for DMD patients.**
3. **Develop a defined local pathway for the cardiac assessment of DMD patients across Wessex.**
4. **Facilitate completion of the Adult North Star Database to develop national standards of care.**

Emerging Leaders Programme
British Cardiovascular Society

## Identifying a Need

Boys are typically diagnosed with DMD by the age of 4 and then receive holistic care in specialist centres directed by paediatric neuromuscular specialists alongside paediatric cardiologists. Locally this is at Southampton Children's Hospital (UHS NHSFT).

Families are typically heavily involved in care and decision making, and rapidly become experts in the disease. Peer support networks are important but can also promote anxiety regarding disease progression due to the variable expression of neuromuscular and cardiac phenotypes.

By the second decade of life most affected individuals are wheelchair bound with very limited skeletal muscle function. Average life expectancy has been historically poor but patients can survive into their 30's and beyond in the era of domicillary non-invasive ventilation to support respiratory function.

Cardiac manifestations of DMD relate to a progressive dilated cardiomyopathy. Most will have developed this by the age of 18. Patients are at risk of clinical heart failure and arrhythmia – including sudden cardiac death (Fig 2).

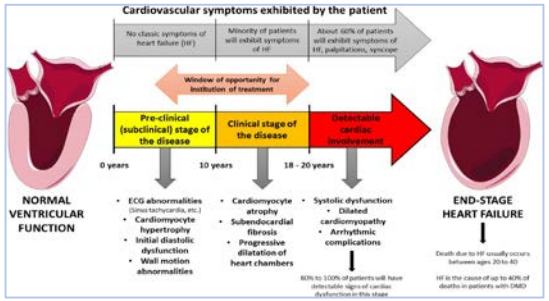


Figure 2: Progressive Cardiac Failure in DMD[1]

Despite all of this, standards of care for Adult patients have been lacking and are only recently published[2]. Cardiac review in adulthood is recommended on an annual basis. To inform practice, the Adult North Star Database has been developed as a multidisciplinary tool to capture the experience of Adult DMD Patients.

Adolescent patients have typically transitioned in an haphazard way with disjointed care of the various aspects of their condition. Cardiology care has historically been transitioned to either the Adult Congenital Heart Team at Southampton or to local cardiology teams. Both patients and staff have found this lack of specialist input concerning.

## Developing the Multidisciplinary Clinic

I recognized that there was an opportunity to improve the experience of Adult DMD patients and their families in the Wessex region following my appointment as a Consultant at UHS.

1. ***Cohorting Patients into Super-specialist Cardiac Clinics:*** DMD adult patients were prospectively identified in collaboration with Paediatric Cardiology and ACHD at UHS and offered initial appointments in specific DMD Cardiomyopathy Clinic slots. Referrals from Neuromuscular Clinic also accepted where adult cardiac care had been delivered at DGH historically.
2. ***Transition:*** As lead for Shared Decision Making (SDM) in Division D at UHS I was made aware of the "Ready, Steady, Go" programme led by the Division A SDM lead and worked to incorporate that programme into DMD care[3].
3. ***Developing Relationships and establishing the MDT:*** I worked closely with the Neuromuscular Consultants locally as well as the specialist neuromuscular physiotherapists for Hampshire and Dorset, the Non-invasive Ventilation Lead Consultant and the Adolescent Palliative Care Consultant to discuss multiple patients. This evolved formally into an MDT.
4. ***Establishing the Multidisciplinary Clinic:*** The MDT worked together to establish a model for the clinic and surveyed patients and carers regarding its acceptability. We identified the regional young adults' hospice Jacksplace as a natural location for the clinic and worked with the management team there to facilitate the clinic as per figure 3 – Healthcare team move between hoist enabled rooms to minimize patient discomfort. I coordinated with our clerical team regarding clinic administration and also with our non-invasive department to provide ECG & echocardiography services utilizing bespoke machines at Jacksplace secured with charitable funding from Muscular Dystrophy UK.
5. **Delivering the Clinic:** The clinic is run quarterly, although the Covid-19 pandemic has interfered with the roll-out. Patients are seen by all members of the MDT except Respiratory medicine due to the requirements for specialist equipment. A formal care plan document is completed as well as the North Star Adult Database as part of an MDT de-brief at the end of the day.

## References

[1] D'Amario D, et al. Heart 2017;103:1770–1779.
[2] R Quinlivan et al. Adult North Star Network (ANSN): Consensus Guideline For The Standard Of Care Of Adults With Duchenne Muscular Dystrophy. Published 6th November 2020
[3] C. F, Biggs et al. Transition of care for adolescents from paediatric services to adult health services. Cochrane Database of Systematic Reviews 2016, Issue 4. Art. No.: CD009794.

## Impact & Experience to Date

Three clinics have successfully run to date. Some patients have attended in-person but others have chosen remote review via NHS Attend Anywhere in the context of Covid19. The MDT have worked together from Jacksplace on each occasion.

Staff have recognized the benefits of direct interaction and the clear advantage of referring concerns to the appropriate service on the same day.

Multiple case histories demonstrate the clinical benefit to patients. Key benefit of introducing Advanced Care Planning.

10 questionnaires back so far. All 10 felt it was beneficial to see all the professionals on the same and all preferred the review at Jacks Places over reviews at UHS.

Feedback:

*"it was much easier having the clinic on one day and easier to get to. It was very beneficial to talk to all the professionals on the same day"*

*"better environment than the hospital"*

*"easier than travelling around hospitals all year"*

*"felt more comfortable and safe rather than going to hospital"*

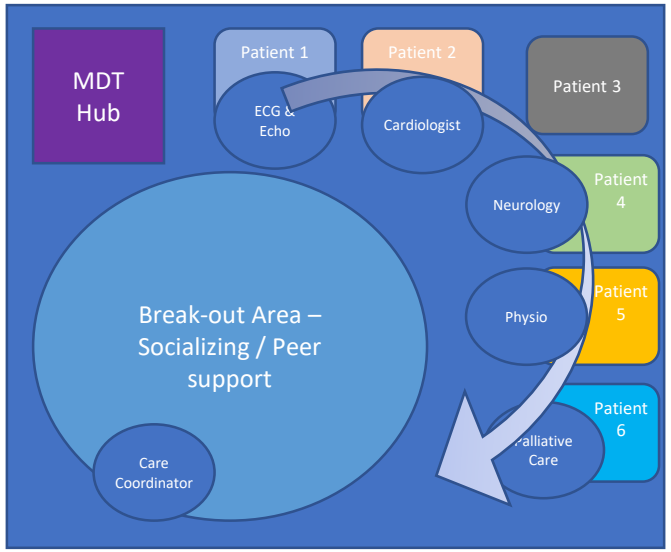


Figure 3. Schematic of the clinic. Patients in hoist enabled, hotel-style bedrooms. Medical staff move between rooms. 1 'rest' station. Breakout area for interaction throughout day.