CLINICAL FEATURE

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Perioperative care planning for a Down’s syndrome patient

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Introduction

This article will be looking specifically at caring for a Down’s syndrome patient, and will focus on the specialist care that they may require throughout the perioperative environment. This article will therefore explain Down’s syndrome and include the related characteristics and health problems associated with this condition, which will then be used to highlight specific areas of care which may need to be improvised in order to accommodate this condition within the operating theatre.

What is Down’s syndrome?

Down’s syndrome is a genetic condition, which is caused by the extra chromosome 21 (Newton 1992). This additional chromosome alters the balance of the body and results in characteristic intellectual and physical features associated with Down’s syndrome. The incidence of Down’s syndrome is approximately one in every 1000 babies born within the UK, and it affects all people regardless of race, age or social situation.

Within a cell nucleus, deoxyribonucleic acid [DNA] strands are divided into small structures, known as chromosomes. Usually there are 23 pairs of chromosomes in each individual, 22 pairs which are identical, and one pair of separate chromosomes, the X and Y, which determines the sex of a new being. The correct amount of chromosome material is necessary in human development. However if this chromosome material is altered in any way, abnormalities are likely to transpire.

The alteration of chromosomal material occurs in a number of different ways, either through non-disjunction, translocation or through mosaicism. These are the three main types of Down’s syndrome; the most common is known as Trisomy 21. Trisomy arises due to a non-disjunction of chromosomes, which is when a pair of chromosomes fails to separate. It results in all cells having the extra chromosome 21, and ensures 47 chromosomes in each cell as opposed to the usual 46. Approximately 94% of people with Down’s syndrome have this type.

Another form of the condition is known as translocation, and accounts for approximately 4% of people with Down’s syndrome. This occurs when a segment of chromosomal material attaches to another in an abnormal way.

The final type of the condition is known as mosaic, and occurs when only some of the cells have an extra chromosome 21, and transpires when cell division goes wrong in the early stages of foetal development. This type accounts for approximately 2% of people with Down’s syndrome (Kessling & Sawtell 2008).

Characteristics of Down’s syndrome

There are many physical features associated with Down’s syndrome. Some of the features are found in the general population, and therefore in order to confirm a diagnosis a Karyotype test would need to be carried out to analyse the chromosomes in the blood cells.

Some of the main features associated with Down’s syndrome include: hypotonia, or reduced muscle tone, flattening at the back of the head, a flattened nose bridge, a small mouth along with an enlarged tongue, small and low set ears, broad hands with a single crease, a big space between the first and second toe, hyper-flexibility of certain joints, reduced birth weight and low set eyes, slanting upwards and with epicanthic folds, which is a vertical skin fold between the inner corner of the eye and the upper eyelid (Rutter 2002).

With regards to mental and intellectual development, children with Down’s syndrome generally progress at a slower rate than other children (Margulies 2007). There is an extremely wide range of ability within Down’s syndrome, although most present with mild to moderate retardation, with IQ’s ranging from less than 20 to over 100 (Cunningham 1988).

Health problems associated with Down’s syndrome

Individuals with Down’s syndrome often have health problems which are commonly associated with the condition. The health issues can range from mild to severe and differ with each person. Approximately 40-50% of people with Down’s syndrome are...
The issues of consent to treatment should also be taken into consideration

born with a congenital heart defect, many of which will require surgery. Upper respiratory tract infections are also common, and result from a deficiency in their immune system. Dry skin, obesity, hypothyroidism and diabetes are also commonly seen with the condition. Eye problems, such as a squint or cataracts are frequently associated with Down’s syndrome, as are dental problems. Gastro-intestinal problems may also present, affecting approximately 10-15% of the population of people with Down’s syndrome (NHS Direct 2008). Leukaemia is also commonly associated with the condition, and develops in approximately one in one hundred children with Down’s syndrome, typically between the age of one and four. Additionally, individuals with Down’s syndrome are at risk of Atlanto-axial instability, which is an increased mobility of the atlas and axial bones, located directly under the skull. This is a potentially critical condition which may result in cord compression, and could consequently result in paralysis (Henderson 2008).

With regards to the ageing process, there is evidence to suggest that some problems related to ageing can occur earlier in people with Down’s syndrome than in the general population. Individuals with Down’s syndrome have a life expectancy of 50-60 years, therefore their health may deteriorate earlier and they are more susceptible to conditions such as depression, dementia or Alzheimer’s disease at an earlier stage in life (Holland & Benton 2008).

The importance of preoperative planning

Care plans are a tool used within the NHS to assess an individual’s plan of care. The preoperative planning assessment is based on individual needs and covers a personalised care plan, incorporating information of health and social care requirements. A personalised care plan is important, and will include details of the services to be provided, an assessment of the individual’s lifestyle and their abilities, treatment plans and care objectives specific to specialities within the healthcare service (Strahan 2008).

Many preoperative assessments are based on a generic care plan, which may not always be suitable for individuals with co-existing conditions such as Down’s syndrome. Appropriate consideration should be given to patients with such conditions in order to prepare not only the individual but also the theatre team in advance of their perioperative care. Many patients with Down’s syndrome like their routine, and coming into hospital can be a frightening time, particularly if they lack understanding about their proposed procedure. This is why it is fundamentally important to ensure that preoperative plans have been discussed and put into place to ensure that their routine is not significantly disturbed, and so that the patient can feel as comfortable as possible within their environment.

The preassessment nurse who is carrying out the preoperative evaluation should have a detailed knowledge of a wide variety of co-existing conditions, including Down’s syndrome, in order to ensure that they follow up and expand on areas of potential complications associated with the condition. For example, if a perioperative care plan does not include information relating to musculoskeletal problems, the preassessment nurse should routinely address whether or not the patient has had any recent changes in motor function or head and neck pain, as this would be important in ascertaining any undiagnosed atlanto-axial instability. These advanced considerations would consequently assist the theatre staff with a documented, quick reference guide and will reduce the risk of potential communication inaccuracies throughout the patient’s theatre experience. It will also assist those involved in the patient’s care with important, relevant and practical pre-considered information, and as a result, will provide best practice in patient care.

Essentially, patients with Down’s syndrome require exactly the same care as everyone else, albeit with a little extra planning. We as professionals therefore need to keep things as normal as possible for them to ensure that they feel secure in their environment, thus supporting a smooth recovery.

Perioperative considerations associated with Down’s syndrome

There are many effects which need to be taken into account throughout the perioperative period when caring for a patient with Down’s syndrome, including both the intellectual and medical implications that may be concerned with each individual.

From a social point of view, it is important for a practitioner to ascertain how much an individual understands about their treatment and care, and consequently communication and interaction with the patient should be altered accordingly. The Health Professions Council Standards of Proficiency for Operating Department Practice states how communication can affect the assessment of patients, and that communication must be adapted to take into account individual factors, such as age or physical and learning disability (HPC 2004).

The issues of consent to treatment should also be taken into consideration. The majority of individuals with Down’s syndrome are considered under The Mental Health Act 1983 (Mind 2006). This act states that those with mental handicap who have enough understanding to give consent for a particular procedure are able to do so, or if this is not the case, consent may be provided by two health professionals, as it is no longer valid in law for a parent or guardian to give consent on behalf of the patient (Newton 1992).

From a medical point of view, it is important to establish an accurate medical history, as there is a possibility that any regular medications could impede any drug therapy. Anaesthetic agents or postoperative pain relief may need to be reconsidered, as the generic choice of anaesthetic induction agent for example may exacerbate any existing conditions such as a congenital heart defect.

The airway will need to be meticulously examined preoperatively, as endo-tracheal intubation may prove to be difficult. It is
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usual for Down’s syndrome patients to have a narrowed airway, a small mouth, an enlarged tongue, enlarged tonsils and adenoids, a high arched palate, a short and broad neck, small or fused teeth, pharyngeal muscle hypotonia which may cause upper airway obstruction, and a small trachea which may require an endo-tracheal tube which is two sizes smaller than predicted (Baum & O’Flaherty 2006).

Patient positioning and manual handling also need to be taken into account. The main reasons for this with relation to Down’s syndrome are for the risks associated with joint laxity and/or atlanto-axial instability. Extra care must be taken to ensure that the limbs and joints are appropriately supported throughout patient transfer and positioning for surgery, and extreme care must be taken with neck support to prevent any cord compression. This also applies to the type and length of surgery the patient is undergoing. Positioning for a tonsillectomy example for example could prove to be incredibly difficult, as hyperextension of the neck should be avoided, again due to the potential atlanto-axial instability, in this particular situation, the patient and operating table should be rotated as a unit, in order to keep movement of the head and neck to a minimum (Baum & O’Flaherty 2006).

Whether the patient will require additional adjuncts such as Flowtron® boots, which may not generally be used for short procedures, should also be recognized and taken into consideration. This care would most likely differ from a theatre’s usual routine for a particular procedure, but may also need to be considered necessary on an individual basis.

Postoperatively, a Down’s syndrome patient would need to be closely observed for airway obstruction. The rationale behind this is that a condition which a Down’s syndrome patient may experience infrequently could present post-operatively and possibly hinder their post-anaesthetic care, for example: sleep apnoea, airway obstruction, or behavioural and/or intellectual problems. Patients with Down’s syndrome are likely to require a familiar face of a parent or carer as they wake up, and therefore arrangements should be in place with regards to the patient’s preference for a particular individual to accompany them in the recovery room. These small, but significant, observations will assist the post-anaesthetic recovery practitioner to provide improved patient care, as they would be presented with an opportunity to ascertain to an extent, the severity of their patient’s condition.

As healthcare professionals, it is important for us to note that individuals with Down’s syndrome are, at some point in their lives, likely to have to undergo some form of surgical intervention. This particular group of patients is more susceptible to infections, which can often have a more severe course and demand of intensive treatment, in addition to requiring medical care from doctors of different specialities.

Whilst there are many sources of information available which describe the potential difficulties surrounding the conditions associated with Down’s syndrome patients, there are very few sources which offer advice on how best to care for such individuals within the perioperative environment. Articles or publications on, or related to this specific topic are not common in professional publications, and some publications such as Health Care Management of Adults with Down’s Syndrome (Smith 2001) are more inclined to focus on the facts of the condition itself, as opposed to offering advice on how best to accommodate or deal with specific areas of care.

Conclusion

In order to excel in the role of an ODP, an enhanced knowledge surrounding the importance and implementation of perioperative care planning is vital, along with an understanding of co-existing diseases and conditions which may influence a patient’s perioperative experience. The overall aim of this article is to bring to light not only the conditions which are commonly associated with Down’s syndrome, but also to emphasize how these conditions may potentially affect patient care within the perioperative environment. As professionals, healthcare practitioners should take into consideration any infrequent situations and be able to accommodate them accordingly.

Each patient has the right to receive high quality care which respects their dignity, confidentiality and equality, and their care should be proficiently communicated to each multi-professional involved within the perioperative environment. We must remember not to stereotype an individual because they have a condition, such as Down’s syndrome. It is the responsibility of each and every team member, within the multi-disciplinary teams, to ensure that the standards of care which are delivered to each patient guarantee that best practice is adhered to. It is important for us to consider that each patient is an individual, and therefore we must treat them as such.

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