Patient experiences of the treatment for acromegaly: the fast track to expert patient

Report from a Focus Group for The Pituitary Foundation

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Introduction
Acromegaly is considered to be a relatively rare pituitary condition. It occurs when the pituitary gland produces too much growth hormone, causing tissues and bones to grow and leading to such symptoms as enlarged hands and feet and changes to teeth and facial features (NHS Choices, 2017). Acromegaly is usually caused by a non-cancerous tumour (called an adenoma) in the pituitary gland in the brain, although in rare cases it can be caused by a tumour in another part of the body (NHS Choices, 2017). It’s commonly reported that three to four people in every million will develop the condition each year, with men and women being equally likely to be affected (Chanson & Salenave, 2008), although this is probably an underestimate as clinical diagnosis of the condition, particularly in the early stages of the disease, is difficult.

Treatment for acromegaly from the NHS depends on the symptoms being experienced. There are a number of treatment goals:
1. Reducing the production of growth hormone by the pituitary gland back to normal levels;
2. Relieving the pressure on brain tissue caused by the adenoma;
3. Treating hormone deficiencies; and
4. Improving the symptoms being experienced by the patient which can include much more than just the changes to appearance detailed above.

Most treatment is likely to involve medication as well as surgery to remove the adenoma from the brain. In some cases medication and radiotherapy may be enough to treat the condition without recourse to surgery. Some individuals will require both surgery and radiotherapy as well as medication before, during and after hospital treatment (NHS Choices, 2017).

The Pituitary Foundation have undertaken two surveys of patient experiences of diagnosis and treatment (Jackson, Murray, Morris & Woods, 2008; Forrest, Norman, Jackson & Percuklievska, 2017). Both reports highlight multiple issues in relation to diagnosis and treatment, and while the more recent of the two reports does include more detailed information for some aspects (such as symptoms likely to be experienced by patients) broken down by pituitary condition, by and large the two reports describe the situation as it relates to the community of pituitary patients as a whole. Our aim in undertaking this work was to gain a better understanding of the needs of patients with acromegaly and the holistic management of their condition in particular.

The survey methodologies employed to date allow for large amounts of data to be gathered, but one drawback when considering rare conditions is whether the questions asked in the survey are adequate to identify the issues of concern for the relevant patient population. Qualitative research accepts that people are complex individuals and we can learn a lot about their experiences and the meanings that they attribute to those experiences by talking to people, asking questions, and allowing their voices to be heard. Focus groups are widely used to explore people’s experiences of, and knowledge about, disease (Barbour & Kitzinger, 1999). As a methodology they provide a forum for the communication of experiences and are particularly suited for this work as they create an environment where issues can be discussed and explored by the participants in a way that perhaps a researcher would not feel appropriate to explore.

Methods
In April 2018 a number of individuals with acromegaly were approached by The Pituitary Foundation’s specialist nurse with an invitation to take part in a Pfizer-funded focus group on the treatment of acromegaly. The focus group took place in May 2018 in a private meeting room at a hotel in the centre of Bristol. The session was facilitated by a research psychologist (SJ, a
member of the Pituitary Foundation Medical Committee), with two members of The Pituitary Foundation staff attending as co-facilitators (GW & RW). The session was videoed and the videographer was present for the duration of the focus group.

(SJ was the focus group facilitator and author of this report, HI nurse, was researcher and patient liaison, with SJ stepping in as lead facilitator due to HI being unable to attend focus group. GW took notes at the event and RW administration for attendees and venue.)

Prior to the focus group, so they could adequately prepare themselves, participants were provided with an information sheet which detailed the aims of the project, the mechanics of the focus group, and details on the topics to be covered in the session which were:

1. Medication
2. Surgery
3. Radiotherapy
4. Symptom management
5. Co-morbidities and associated risks

Informed consent was taken from the individuals prior to the start of the focus group. Seven people with acromegaly took part in the focus group which lasted for approximately 2.5 hrs. Group members’ ages ranged from their mid-20s to mid-60s; four were female, while three were male. Time since diagnosis varied widely – some relatively recent, while for others diagnosis had been over 30 years prior. All members of the group had required treatment for acromegaly but those treatment experiences varied widely.

**Results**

We have tried to reflect the range of treatment experiences reported by the participants. It should be noted that over time the management and treatment of acromegaly has changed, so treatment options for older members of the group were not the same as those for younger members of the group.

Themes arising from the discussion of the topic areas have been identified and agreed between SJ, GW & RW. The themes are described by topic area and anonymized quotes from the focus group participants are shown in italics. Although not mentioned by the participants, comparisons are made with the information supplied on the NHS Choices website as this is a likely source of information for individuals with acromegaly.

The first three sections of the results (medication, surgery, and radiotherapy) were each discussed as separate topics in the focus group. The final section (living with symptoms, co-morbidities and associated risks over the longer term) reflects the fact that these issues were discussed together in the focus group. For each of the sections detailed in the results given below a video extract from the focus group is available on The Pituitary Foundation website.

1. **Drugs**

The NHS Choices website tends to suggest that medication may be prescribed after surgery if the levels of growth hormone produced by the pituitary gland have not returned to normal (NHS Choices, 2017). However, it was clear from the experiences related by the members of the focus
group that drugs are used at all stages along the patient journey for different reasons at different times.

For example, pre-surgery drugs can be used to either shrink the tumour or to reduce growth hormone levels. One participant had been given bromocriptine while two had had cabergoline prior to surgery – both are drugs intended to stop growth hormone being produced, however, they don’t work for everybody. Like all drugs cabergoline has side-effects, although for one of the participants this seem related to dosage. At lower doses there were no side effects, but at higher doses she experienced the feeling of a racing heart. For the other participant side effects included headaches, nausea, extreme mood swings, depression and mild psychosis all of which improved when the drug was stopped. One of the participants was also prescribed lanreotide prior to surgery, one of a trio of drugs (including octreotide and pasireotide) usually administered in the form of a monthly injection which aim to slow down the release of growth hormone and which can sometimes shrink tumours. For the participant the lanreotide caused diarrhea which meant “couldn’t go out on the weekend following the injection. Didn’t bring down the growth hormone or the size of the tumour.” Another participant, prescribed lanreotide after surgery, had no side effects and was chosen by the HCPs for ease of administration and probably because it was cheaper than octreotide, however, while it might be cheaper it didn’t work to reduce the growth hormone levels even when the injection schedule was modified to be every three weeks rather than monthly, although the change in schedule did help with the pain and fatigue management. One participant’s experiences with Somatostatin LAR involved having to tell the nurses how to mix the medication for administration which could have been stressful and worrying.

Some participants had experience of drugs being used as part of post-surgical treatment. As with the situation described pre-surgery, participants tried different drugs with mixed results. Octreotide in tablet form was associated with minimal side-effects for one participant, but the switch to the monthly injection was less successful in controlling the growth hormone which meant a change to a different drug, pasireotide, which “nearly took me into diabetes” and as a consequence had to be stopped. Only one participant had been given pegvisomant which effectively reduced growth hormone levels and had few side effects other than weight gain for most of the time. However, after taking the medication for some time an increase in levels of liver enzymes meant stopping for a while and restarting at a lower dose.

All the male participants required testosterone replacement post-surgery. Testosterone replacement comes in a number of different forms and it’s another example of trial and error finding something that suits each individual. For example, with the injections there is an initial peak but as it wears off tiredness increases and beard growth slows. This is not the case with a regular low dose, but not everyone likes the gel formulation that the drug comes in to enable that and “with the injections you can have it and then forget about it for 10/12 weeks”.

So at whatever point in the patient journey medication is used there always seems to be an element of trial and error - trying different drugs at a variety of dosages and in different forms. Patients have to achieve a trade-off between the effectiveness of the drugs and being able to manage the side-effects, while still managing activities of daily living as well as having some quality of life. There are some additional practical matters that also have to be considered, for example, the storage of the drugs, and planning food consumption. Participants understandably have concerns about using drugs over the longer term.
If part of being an expert patient is being able to communicate effectively with HCPs about treatment and making treatment choices, then with acromegaly this seems to begin fairly early in the patient journey. None of the members of the focus groups spoke about any problems in discussing different drug options with HCPs, although it did sound as though HCPs are more likely to suggest the cheapest most suitable drug options first, moving up to the more expensive ones as it proves to be necessary to do so.

2. Surgery
There is limited information on the NHS Choices website about surgery for acromegaly. It provides only a brief description of the surgical process to remove a pituitary tumour as well as a bullet-pointed list of the risks associated with such surgery which should be discussed with the surgeon prior to treatment (damage to the healthy parts of the pituitary gland, leakages of the cerebrospinal fluid (CSF), and meningitis) (NHS Choices, 2017).

The group were clear that it is very important to have a full list of questions to ask your surgeon prior to the operation to help you prepare properly (“don’t be scared to ask”). As the condition is rare, finding a surgeon with enough experience is important and will involve talking to other people and asking consultants and HCPs for advice and recommendations. Travelling to specialist centres isn’t possible for everyone, in which case you may have to accept being treated by a less experienced surgeon and nursing team in a more convenient location which one group member described as “stressful”.

All the members of the focus group had undergone surgery as part of their treatment for acromegaly, four of the group having one surgery, the remaining members of the group requiring three operations each. Experiences ranged from being very straightforward with no complications of any kind, to very complex with multiple surgeries and other treatments required to address the various complications that had arisen. The complications experienced by the group involved CSF leaks, meningitis, hyponatremia (low sodium levels in the blood), septic leg (from graft site to fix CSF leak becoming infected), carotid artery repair, and developing diabetes insipidus. What was most striking was how much of what happened to the group members was unexpected which doubtless contributed to the stress and worry of what they experienced.

If we take the CSF leak as one example of a post-operative complication - a CSF leak is considered to be a fairly rare complication arising from the surgery, however, it occurred for five members of the group. What is striking within the group is the unexpected variety of experience related to this one complication. One member of the group developed a CSF leak during surgery, the person was very surprised to wake up and find that a graft had been taken from his leg to repair it. “Had not been made aware of this possibility prior to surgery.” Another member of the group required complicated surgery to remove the tumour. The surgery was successful but they developed a CSF leak post-surgery whilst still in hospital, and didn’t know what it was: “Was sent home with a drip, knowing very little about what it was. Didn’t query it.” After becoming increasingly ill at home he ended up being taken back to hospital with viral meningitis which involved various treatments during an extended stay in hospital. The other member of the group who was sent home with a CSF leak said it did eventually stop “but it was an extremely worrying time for me and my family. My mother had to watch me sleeping every night as patients have to sleep sitting up.” Two members of the group also developed the CSF nasal drip between two and five weeks post-surgery. In one case, the hospital’s response was to ask her to send them a sample of the fluid in the post for them to test while the staff at her local A&E department lacked the knowledge to diagnose her. On her return to hospital, fixing the CSF leak took two attempts in
part because the first graft using abdominal tissue failed. “Left hospital 3 weeks later extremely nervous it hadn’t worked again and felt incredibly scared. A horrendous experience.” For the other lady, the HCPs told her it wasn’t CSF but she was eventually rushed back to hospital and into theatre for more treatment.

There are a number of possible complications and problems that can lead to unexpectedly long stays in hospital. It seems that patients with acromegaly have to become expert patients relatively quickly, developing the confidence to explain their symptoms and concerns and request treatment. This seems to be particularly the case in relation to CSF leaks where it feels the onus is very much on the patients to go into surgery very well informed as a CSF leak may develop at a number of time points, it carries a significant risk of making individuals very unwell and HCPs, especially those who work in more general areas of medicine, may not recognize either the condition or the risk.

There was general agreement that everyone’s acromegaly journey is different, and that “it’s important to put forward a balanced view to patients needing surgery.” Especially important in these days of social media where “people on forums tends to be the people who have had bad experiences” which they need to share but where the voices of those for whom things have been straightforward tend to be missing.

3. Radiotherapy

The NHS Choices (2017) website says that radiotherapy may be offered in cases where surgery isn’t possible or wasn’t totally successful in removing the tumour, or where medication isn’t working. The stated aim of radiotherapy treatment is to reduce growth hormone levels, and there is a warning that its effect may well not be immediate. There is a description of the two types of radiotherapy used to treat acromegaly (stereotactic and conventional radiotherapy). There is a brief description of the possible side effects of the treatment on hormone levels generally and on fertility, with a note that you should discuss the risks and other possible side effects (not identified) with your doctor.

While the treatment itself is relatively straightforward although quite intensive, patients need to be convinced that it’s needed possibly because of “fear in the media of possible long term effects”. Being properly prepared for the experience is very important. A lack of proper preparation in terms of knowing what to expect can result in a fairly traumatic experience. For example, one lady was inadequately prepared for the table to move and spin during the planning session which she found so difficult to cope with she had to ask the staff to stop the session.

Radiotherapy may be delivered as a course of treatment over a period of weeks which can be associated with a feeling of fatigue or short term memory problems. All the members of the group were clear about “the need to pace yourself. There are up and down periods in your recovery.” Getting into a routine for the duration of the treatment helps, and “remain positive”. This is the element of treatment for acromegaly where individuals seem to make the most direct comparisons between themselves and patients with cancer. Both acromegaly and individuals with cancer can require radiotherapy, but if you have acromegaly there can be a sense of guilt at struggling with the treatment and needing support to deal with it.
There were some concerns about the long term effects of radiotherapy such as unremitting fatigue and the need for increasing amounts of differing kinds of hormone replacement therapy over time.

In contrast to surgery, radiotherapy is seen as being relatively straightforward, not least in part because it seems to take place further along in the patient journey when the patient has amassed greater knowledge about the condition and its treatment. That said, good quality information to enable patients to be well-prepared is key and not always readily available from HCPs.

4. Living with symptoms, co-morbidities and associated risks over the longer term

The NHS Choices (2017) website says that treatment is often effective but recognizes that there will be a need for life-long monitoring with two main aims:

1. To make sure that any hormone replacement is adequate; and
2. To make sure that the condition hasn’t returned.

Once the acute phase of treatment is over the monitoring begins. Focus group members talked about what it felt like to be considered “cured” or “in remission” by HCPs, themselves and others. “Being classed as in remission is difficult because it feels like it could always come back.” Some group members felt that HCPs “don’t take you that seriously when you’re classed as being in remission.” One group member said his endocrine team class him as “a maintenance project”. There was a feeling that the HCPs tend to “look at the data and not at how you’re feeling.” One of the group reflected that she feels “her remaining symptoms are now overlooked” with endocrine appointments focused purely on medication and test results. There was a clear sense that by this stage, patients have “become experts in our own condition and our bodies”. With the acknowledgement that some HCPs (particularly doctors) are quite unlikely to treat this stage holistically, there was a sense of a need to deal with the situation proactively: “we need to start telling them [the HCPs] how we feel and not wait to be asked.” This includes insisting on continuity of care which was seen as being very important.

The uncertainty about what might happen in terms of longer term effects of treatment (especially radiotherapy) places a considerable psychological burden on patients. “The tumour may have gone but the baggage continues for many years.” “Coping with recovery means you don’t know what’s round the corner.” Symptom management is a significant focus post-surgery – the group members between them had quite a list of issues to deal with including: hypopituitarism, extreme fatigue, weight gain, joint pain and problems with knees and hips, short term memory problems, early menopause, infertility, cardiovascular problems, visual problems, problems with gum disease and teeth, enlarged tongue, and no/heavy periods.

There was a strong feeling on the part of the group that the psychological aspects of acromegaly are neither well understood nor well supported. “The changing of the self can be psychologically damaging.” “The psychological journey has had the biggest effect ... it has taken about 18 months to get some sort of confidence back.” For men in particular the problems around the effects on their sex drive with knock-on effects to their sex life is a very big issue isn’t acknowledged or discussed enough. The appearance changes associated with acromegaly pose a significant psychological challenge regardless of gender: “the psychological damage of your physical change is the worst thing for acromegalics. You no longer look as you perceive yourself to be.” Talking
about the changes to her head, face and hands, one participant reported considerable anxiety and went so far as to describe herself as “a monster”. While one of the male participants reflected that it took him 10 years to feel comfortable with his appearance again.

The symptoms and the psychological impact are difficult to deal with and to explain to others (including HCPs), and can leave people feeling “incredibly isolated as it’s a rare condition … [you’re] alone dealing with things people are not aware of.” That said, there was also a sense that acromegaly “empowers you to become an advocate for the condition”. And, “if you can get through it, then you can help others to do the same”. The role of The Pituitary Foundation in providing support and information was acknowledged as invaluable, but there’s a continuing need to keep getting the message out about it.

The drugs, surgery and radiotherapy offer a cure, but at what cost? Acromegaly needs to be caught quickly and treated early to have the best possible outcomes for patients. While it is possible to gain remission from the tumour through treatment there are likely to be some long-term effects which can be difficult to live with, such as problems with sexual function, reduced quality of life, feeling isolated and lonely. It can be hard to lead a normal existence with such long-term effects to deal with.

Conclusions:
The information about the treatment options on the NHS Choices website is potentially misleading. Acromegaly is a rare condition, and the experiences of the members of the focus group indicate that each person is likely to need the treatment tailored to meet their particular biology, physiology and circumstances. As far as medication goes, a certain amount of trial and error is needed with regular monitoring in case symptoms change. It is clear that surgery can be both a worrying and frustrating experience in part because it happens fairly early in the patient journey at a point when the individuals are still learning about their condition. While radiotherapy is considered a more straightforward treatment it is still a worrying procedure to undergo and good preparation for it is important. A lack of knowledge in respect of any of the treatment areas can leave patients quite vulnerable and dependent on healthcare professionals who do not always communicate very effectively. Over the longer term, patients have to find ways to motivate themselves to make the most of the good times when they present themselves as the on-going symptoms introduces an inescapable current of uncertainty reinforced by the regular monitoring. There is a degree of frustration that long-term monitoring on the part of healthcare professionals tends to focus on blood and MRI results rather than any attempt to elicit from the patients themselves the more holistic and psychological issues that may need to be addressed.

Learning points

- The information provided on the NHS Choices website is far too limited to be of real practical use for patients preparing for treatment for acromegaly. Encouraging the website owners to add a link to The Pituitary Foundation website would be a useful addition.
- Treatment can be very simple and straightforward, but it can also be complicated. Patients need access to good quality information both to reassure them but also to prepare them to know what to look out for and raise as concerns with healthcare professionals. The Pituitary Foundation has a number of useful booklets and fact sheets that HCPs could refer patients to.
- The development of a patient passport for use by diabetes patients has improved their hospital healthcare experiences, and a similar document for patients with acromegaly
would potentially enable patients to communicate with more confidence and authority when receiving treatment.

- Promote the use of the Pituitary Distress Thermometer as a way to empower patients to identify and discuss the wider issues associated with acromegaly with HCPs.
- More awareness raising is needed to make more HCPs aware of the existence of The Pituitary Foundation and the services it offers to patients with acromegaly.

**Resources**

Acromegaly Focus Group videos on the Pituitary Foundation website


Pituitary Foundation acromegaly booklet – covers symptoms, diagnosis, treatment and surgery

Pituitary Foundation Pituitary Surgery & Radiotherapy booklet – provides in-depth information on what to expect

Pituitary Foundation Post-Surgery Fact Sheet – provides information on what to expect and when to seek help

**References**


