

Anaesthesia recommendations for **Xeroderma pigmentosum**

Disease name: Xeroderma Pigmentosum (XP)

ICD 10: Q82.1

Synonyms: Kaposi disease, Ichthyosis; individuals suffering from this disease are often referred to as children of the night or moon people

Disease summary: Xeroderma Pigmentosa (XP) is a rare autosomal recessive disorder due to the defect in the nucleotide repair genes resulting in inability to repair the damaged DNA caused by Ultraviolet (UV) rays. Hence people suffering from this disease show extreme sensitivity to sunlight and UV radiation and are characterized by skin lesions mostly in the sun exposed areas such as head, face and neck. It affects 1 in 2.5 million patients and has 1000 fold increased risk of developing cancers in sun-exposed areas. Even the incidence of internal malignancy is 10-20 times higher than in normal individuals. Disease may also be associated with progressive neurological degeneration. Most people require multiple surgeries for removal of the skin, ocular lesions and malignancies. The main anaesthetic concerns are difficult intravenous cannulations, difficult airway (bag-mask ventilation and intubation), genotoxic and progressive neurological deterioration of the patients with the use of volatile anaesthetic agents, increased sensitivity to opioids, benzodiazepines and muscle relaxants, and difficult extubation due to epiglottis subsidence.

Medicine is in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong



Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

Typical surgery

Patients with XP undergo excisions of Squamous cell carcinoma, Basal cell carcinoma on the UV exposed areas (face, head, neck) along with skin graft and skin flaps.

They also undergo eyeball enucleation, ectropion correction, eye tumours removal, and cataract surgery.

Type of anaesthesia

The lesions are usually present in the head, face and the upper part of the body, so most of the patients require general anaesthesia.

Volatile agents should be avoided as these agents have been shown to facilitate the progression of the disease. Halothane has genotoxic effects and Isoflurane, Sevoflurane are associated with worsening of neurological symptoms. So total intravenous anaesthesia should be the preferred anaesthetic technique.

Such patients are sensitive to benzodiazepines and opioids. Thus premedication with Benzodiazepines are better avoided and if necessary, such drugs should be administered under monitoring.

These patients are more sensitive to muscle relaxants. So total avoidance of muscle relaxants if possible, and if not then at least a smaller dose of shorter acting muscle relaxant, with strict neuromuscular blockade monitoring is recommended.

Whenever possible, if the surgical site permits, regional anaesthesia should be preferred over general anaesthesia

Necessary additional pre-operative testing (beside standard care)

Genetic screening can be done to know the type of XP (total 7 types, XPA to XPG) and also to know about the neurodegenerative type (all are, except for XPC and XPE).

Neurological assessment as well as CT/MRI of the brain in the patients having neurological symptoms to rule out any causes other than XP.

Sensorineural hearing deficiency tests in patients having hearing loss.

Particular preparation for airway management

There are cases reported to have difficult airway in patient suffering from this disease. Multiple lesions on the face and facial disfiguring can cause difficult face mask ventilation, thus multiple size and types of face masks should be prepared accordingly.

Also, there are reports of difficult intubation requiring stylet and bougie during intubation. So difficult airway cart should be available during airway management. Awake fiberoptic intubation can be the choice of airway management in patients with anticipated difficult airway.

We also advice to keep gauze piece soaked with paraffin in cases of ulcerative lesions to prevent bleeding and peeling of skin from direct pressure of the mask.

We also suggest awake extubation in patients with epiglottic dehiscence, which are reported to cause postoperative stridor.

Particular preparation for transfusion or administration of blood products

No recommendations found.

Particular preparation for anticoagulation

No recommendations found.

Particular precautions for positioning, transportation and mobilisation

Disease progresses when the skin is exposed to the UV rays. So patient should wear protective clothing, UV shielding films and sunscreens having high sun protective factor (SPF) in the operation theatre.

Covering all the parts of body with proper drapes before turning the operating room lights on and during whole of the intraoperative period is advised. Avoidance of halogenated lights as much as possible should be done.

Bones may be fragile in these patients due to avoidance of sunlight leading to Vitamin D deficiency, so proper padding of pressure points and gentle handling during transport is a must.

Use of eye ointments and proper closure of the eyes must be ensured in patients having conjunctival and corneal lesions.

Interactions of chronic disease and anaesthesia medications

The patients may be taking chemotherapeutic agents like 5-Fluorouracil (5-FU) which causes myelosuppression. So nitrous oxide should be avoided in such patients.

Patients with XP may be on steroid therapy, so steroid supplementation in the intraoperative and maintenance in the postoperative period is advised.

Anaesthetic procedure

Due to multiple lesions on the skin, intravenous cannulation and fixation can be difficult.

For general anaesthesia total intravenous anaesthesia (TIVA) is preferred. Halothane, Isoflurane, and Sevoflurane have shown to have genotoxic effect and also have caused

worsening of neurological symptoms. So these agents are better avoided. Propofol, Ketamine or dexmedetomidine with short acting opioids are the preferred anaesthetic agents for TIVA.

Nitrous oxide should be avoided in patients taking 5 Fluorouracil as both the drugs cause myelosuppression.

Since these patient are sensitive to the paralyzing effects of muscle relaxants due to neuronal dysfunction and muscle atrophy, total avoidance or minimal use of shorter acting agents with strict neuromuscular block monitoring is advised. Intubation without the use of muscle relaxants, can be facilitated by the use of other anaesthetic adjuncts like dexmedetomidine.

Besides, Dexmedetomidine also reduces the induction and maintenance dose of propofol, blunts the airway reflex during intubation and extubation, decreases the requirement of opioids and also helps to enhance the recovery. Ketamine can also be a good adjunct of TIVA.

Also, these patients are very much sensitive to opioids. So short acting opioids such as fentanyl can be used for the intraoperative period. Use of multimodal analgesia with ketamine, paracetamol, ketorolac, dexmedetomidine, infiltration of local anaesthetics decreases the requirement of opioids. Concomitant use of benzodiazepines and opioids have led to apnea and desaturation indicating that these patients are very sensitive to this combination.

Eye ointment use in the intraoperative periods prevents the drying of cornea and thus, corneal ulcerations.

There is no contraindications for regional anaesthesia provided by the site of the surgical field and patient's choice.

Particular or additional monitoring

Standard ASA I and II monitoring is required for all patients which include temperature, heart rate, ECG, Blood pressure, peripheral oxygen saturation concentration and end-tidal carbon dioxide concentration.

Peripheral nerve stimulator to assess the neuromuscular blockade is recommended to be used when muscle relaxants are used.

Bispectral Index (BIS) monitoring is beneficial to ensure the depth of anaesthesia where ever applicable.

Possible complications

Progression of the disease and worsening of the neurological symptoms in the immediate and late post-operative period occur in patients especially when volatile agents such as Halothane, isoflurane, sevoflurane is used.

Delayed recovery, agitation, abnormal movements, confusion are the immediate manifestations whereas memory disorders, false recognition and reversible cognitive decline have been reported as the late complications after the use of volatile agents.

Post-operative care

Avoid the exposure of the skin to UV lights in the exposed parts.

Monitor for the smooth recovery as drugs effect are more pronounced in such patient.

Disease-related acute problems and effect on anaesthesia and recovery

No recommendations found.

Ambulatory anaesthesia

No recommendations found.

Obstetrical anaesthesia

No recommendations found.

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Please note that this recommendation has been reviewed not by an anaesthesiologist and some other disease expert but by two anaesthesiologists instead.
